PEDIATRIC CARDIOLOGY

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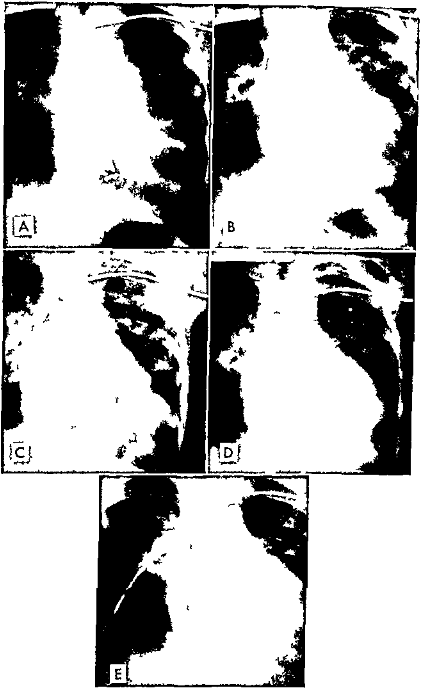


Figure 115 A Cardiac catheter approaching from the left arm and reaching the border between innominate vein and superior vena cava B Cardiac catheter in the midlateral portion of the right atrium C Catheter advancing through the tricuspid valve into the right ventricle D Catheter entering pulmonary artery from the right ventricle E Catheter in the pulmonary capillary position wedged out into the lung fields

maneuvering the catheter cannot be advanced in the required fashion and the study cannot be completed While the tube is being advanced the patient is connected to a direct writing electrocardiograph or oscilloscope and one member of the team watches the tracing for the appearance of arrhythmias

When the catheter reaches the superior vena cava (Fig 115 A) the saline drip is disconnected while 3 or 4 cubic ml of blood are collected in a heparinized syringe for oxygen analysis This is handed to a technician who places it in an ice bath The drip is then reconnected and the catheter is advanced into the right atrium where another blood sample is obtained (Fig 115 B) After this has been accomplished the outlet of a manometer is attached to the catheter and a right atrial pressure tracing is obtained Then the catheter connected to the recording apparatus and under fluoroscopic control is advanced from the right atrium to the right ventricle which is identified both by the appearance of a ventricular type of pressure tracing and by the position of the tip of the catheter under the fluoroscope (Fig 115 C) A blood sample is obtained from the ventricle then under fluoroscopic and manometric control the tip is advanced into the pulmonary artery (Fig 115 D) A blood sample having been obtained here the catheter is again advanced as before and wedged as far out to the lung fields as possible into a so called pulmonary capillary position where samples and pressures are again obtained (Fig 115 E) When all this is completed the catheter is pulled back into the pulmonary artery It is left here while a timed sample of expired air is collected into a Douglass bag an arterial blood sample is withdrawn and a pressure tracing is obtained from the brachial artery through the "cut down" Blood samples and pressure tracings are obtained in identical fashion from the right ventricle right atrium and superior vena cava again on the way out The catheter is removed and the "cut down" is sutured

The entire procedure from "cut down" to suture closure usually takes one to two hours In our experience a general anesthetic is required only rarely Morphine and barbiturate sedation serves the purpose adequately in most instances Exposure to fluoroscopy should last no more than fifteen minutes

As may be seen from the description at least four people are necessary for the efficient performance of the procedure one handles the catheter one is in charge of the pressure recorder and watches the electrocardiogram a third handles the fluoroscope and a technician takes care of the blood samples In addition to these four a nurse preferably one familiar to the child should also be present Rarely the services of an anesthetist are required

In addition to the fluoroscope the sterile surgical instruments and the catheter an electrocardiograph and one or two manometers connected with a multichanneled recording apparatus are necessary A

Foreword

Sixty years ago Dr Thomas Morgan Rotch then Professor of Diseases of Children published the first textbook on Pediatrics from the Harvard Medical School Even in 1896 there was enough information regarding this relatively new specialty to require a volume of some 1100 pages but only seven of these were headed Congenital Diseases of the Heart Obviously there was relatively little to say about this particular pediatric problem and even less stimulus toward further knowledge for Dr Rotch wrote that although "it is usually possible to make a diagnosis of congenital cardiac disease a diagnosis of the especial lesion is as a rule impossible" In any case the exact diagnosis was only of academic interest In his single paragraph devoted to the management of con genital heart disease Dr Rotch did mention "the administration of digitalis in small doses and with the utmost caution" as occasionally useful but concluded that the treatment was "essentially hygienic and symptomatic"

The next textbook of Pediatrics from the Harvard Medical School appeared thirty years later Professor John Lovett Morse was able to compress what he thought the pediatrician of 1926 needed to know about heart disease into forty pages less than five of which sufficed for Con genital Heart Disease Doctor Morse could find one item of progress unknown to Dr Rotch But even this was nearly useless for "the Roent gen ray which theoretically ought to be of considerable assistance in the diagnosis of special lesions is practically of little assistance even in the hands of an expert Yet "Fortunately the diagnosis of the exact lesion is not of great importance in either prognosis or treatment There is no curative treatment [and] nothing which will either diminish the deformities or favor the closure of abnormal openings The treatment must therefore be hygienic and symptomatic"

Now in 1956 we find ourselves at the end of another thirty year interval Judged by the progress before 1926 the mere size of this new book on Pediatric Cardiology from the Harvard Medical School and Childrens Hospital of Boston is sufficiently impressive Even more

striking is the preponderance of pages devoted to the diagnosis and treatment of congenital as compared with acquired heart disease Obviously the sudden increase in knowledge and interest concerning a subject until recently so neglected is worldwide Its local history has been traced here only as a convenient example of the size and suddenness of this development Why should one thirty year interval (indeed essentially the last fifteen of those thirty years) have witnessed such striking progress?

A partial explanation is found in the decline of other challenges such as the feeding methods formulas and digestive diseases of infancy and childhood But the real cause was the demonstration by Gross in 1939 and by Blalock and Taussig in 1945 that it was indeed possible to diminish the deformities or favor the closure of abnormal openings The essential element was some basis for hopefulness With hope interest in diagnosis came suddenly to life cardiac catheterization became a useful tool rather than a *tour de force* "the Roentgen ray" was belatedly found to be of considerable assistance indeed and the "symptomatic and hygienic treatment took on entirely new meaning and purpose

The pediatrician—and especially the younger pediatrician—has rightly been unwilling to see all these aspects of diagnosis and treatment handed over to a new subspecialist in cardiology Fully aware that the surgeon who operates upon the heart of a child requires special skill and training the pediatrician considers the nonsurgical aspects of cardiac disease in infancy and childhood to be pediatric responsibilities In rising to meet this new and growing challenge he will find this volume extremely helpful Its author once described to me his hope of making this "a do it yourself cardiology" I believe he has succeeded

Doctor Nadas is especially well qualified for this task His approach as an author parallels that of his own professional life His residency in Pediatrics at The Childrens Hospital in Boston and his year of full time pediatric responsibility and teaching at the Childrens Hospital of Michigan in Detroit were followed by four years of general pediatric practice in a small city some distance from the specialized services of a medical school With this background of practical experience Dr Nadas returned to The Childrens Hospital in Boston in 1949 to participate in the suddenly and rapidly expanding activities of its Cardiology Division Now as Cardiologist to the group of hospitals making up The Childrens Medical Center and Consultant in Pediatric Cardiology to the Boston Lying in Hospital the author would I believe prefer to be known primarily as a thoughtful and resourceful doctor for infants and children That Dr Nadas is also a wise specialist of large experience in cardiology and a teacher of great skill and enthusiasm will be abundantly clear to the reader of the following pages

CLEMENT A SMITH MD
Department of Pediatrics
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Preface

This volume is intended to be a handbook for the pediatrician, the general physician and the medical student. It does not claim to be a treatise on the physiology of the heart, though obviously physiological data will be presented in an attempt to explain the clinical picture. I have tried to make every chapter fairly complete by itself; thus a certain amount of repetition is unavoidable.

My intention has been to put proper emphasis on clinical recognition and management of heart diseases in children. Techniques of cardiac catheterization and angiocardiography, for example, have been described, but primary emphasis has been kept on the principles and procedures of clinical management.

I have written almost exclusively about our own experiences at The Children's Medical Center during the past ten years, and I did not intend to write a reference book. Diseases and even important therapeutic procedures, such as open heart surgery and hypothermia, with which I had little personal contact, have been discussed only cursorily. The interested reader may do better to look up these topics in the reference material rather than have me interpret other people's work for him. The list of references, like the book itself, is a subjective one. I did not aim at completeness, but rather included only the books and articles I have found particularly interesting and useful through the years.

The list of people to whom I am indebted—directly or indirectly—in helping me complete this work would easily fill a whole page.

I want to start with my mother and father. The memory of my father, a writer who always wanted to be a doctor, inspired me particularly. I felt that I was paying homage to him by fusing in this volume the profession of the writer with that of the physician. The little emblem at the foot of this preface was the trademark of his magazine published many years ago in Budapest, Hungary.

On the professional side I should like to begin with Dr. Charles A. Janeway, Thomas Morgan Rotch Professor of Pediatrics at Harvard Medical School, who gave me the opportunity to work in the field of

pediatric cardiology Doctor Clement A Smith friend and teacher has helped me in many ways during the past fifteen years I want to express my sincere thanks to Drs Edward B D Neuhauser and Martin H Wittenborg who taught (and fought) me in radiology and to Dr Robert E Gross for much invaluable teaching and the use of his patient material Doctors Abraham M Rudolph and Walter T Goodale my associates in the laboratory assisted me in collecting and interpreting the physiological data presented Without the help of Drs Patrick A Ongley and Donald C Fyler I am sure the illustrations would not have been completed in time I would also like to thank Dr Robert M Smith for his contribution in writing the section on Anesthesia

I would like to express my sincere appreciation to Mr Walter T Hall who through the establishment of a fund for the support of research in pediatric cardiology helped to defray expenses of publication

I wish to thank my secretary Miss Myrtle R Westhaver for her efficiency in holding together the cardiology department during the trying period in which this book was written Mrs Marcia Lawson for the editing of the manuscript Mr Ferdinand R Harding for the photography Mrs Margaret M Steele for the typing and Miss Mary E Delaney for the illustrations

Finally I want to thank sincerely the W B Saunders Company for the many patient ways by which they helped me prepare this volume and efficiently bridge the Atlantic with their galley proofs

ALEXANDER S NADAS M D

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January 1957

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Part Two ACQUIRED HEART DISEASE

PART ONE

THE TOOLS OF DIAGNOSIS

CHAPTER 1

History, Physical Examination and Routine Tests

HISTORY

The history in pediatric cardiology should encompass the principal pediatric and cardiac features of the disease.

The history should begin with a résume of the chief complaints. It is extremely important to learn right from the outset how sick the child really is and how many of the complaints are entirely iatrogenic in origin and secondary *only* to the discovery of a heart murmur. The entire picture in a child who is sick because the doctor said so, who is "too active" and whose limitation of exercise is the result of parental restriction will be different from that of *another* youngster who "will not gain or grow like others, gets blue, or breathes heavily."

If the presence of a murmur represents a significant portion of the complaint, it is vital to elicit certain facts regarding its timing and the circumstances of its discovery. Was it present at birth? If not, how long after birth was it discovered? Was is discovered by a physician who had been taking care of the youngster all along or by a new doctor who was seeing him for the first time? How closely had the child's health been supervised prior to the discovery of the murmur? Were there febrile illnesses, especially sore throats, immediately preceding its appearance and, if so, what was the time interval between the occurrence of the sore throat and the appearance of the murmur? Precise information related to these points may on occasion be the only clue on which to base a differential diagnosis between congenital and acquired heart disease.

Next, the history of the present illness should be elicited. This should be a detailed chronologic account of the child's disease from the first time his health was questioned. It is important that the inquiry in regard to the present illness should not reach back only to the time when heart disease was first diagnosed. In many instances, especially in cases of acquired heart disease, cardiac disorder or its significant antecedents may have been present before there was any clear indication of involvement of the circulatory system.

A careful systemic review is perhaps the most important part of the history In regard to the gastrointestinal system the question of appetite should be raised if the appetite is poor especially in an infant one ought to inquire whether the mother believes this may be due to fatigue or whether it represents a general lack of interest in food Parents should be questioned carefully concerning weight gains and losses A history of abdominal pain may raise the suspicion of rheumatic fever The genito urinary system should be considered particularly in respect to a decrease in urinary output characteristic of congestive failure In adolescent girls especially cyanotic ones the effect of the onset of menses on general well being and exercise tolerance should be questioned

The cardiorespiratory system obviously is the most essential part of the systemic review An excellent idea of the exercise tolerance of these patients may be obtained by careful questioning about the average distance they can walk on level ground or the flights of stairs they can climb without undue fatigue One inquires into their playing habits and how they keep up with their contemporaries In infants and small children who do not walk or run the effort of taking a bottle of sitting up for a definite length of time or of crawling around the playpen may represent a rough test of exercise tolerance One seeks information regarding dyspnea or wheezing The child s breathing when he is asleep is a particularly important criterion of respiratory effectiveness one should note the rapidity of respirations the use of ancillary muscles (alae nasi) and the presence or absence of grunting A vivid description of the forcefulness of cardiac action can sometimes be obtained by asking parents whether they themselves could feel the child s heart beating against the chest wall The time of onset of cyanosis its permanent or paroxysmal nature its severity progression and localization are all important data One should inquire about the presence or absence of squatting habits and also other favorite positions (such as knee chest) which the child is likely to assume when tired Such positions strongly suggest a cyanotic type of congenital heart disease particularly tetralogy of Fallot The number type and severity of respiratory infections through the years should be established these are usually increased in patients with large left to right shunts and they may predispose to rheumatic fever Inquiries should be made regarding chest pain and hemoptysis particularly in older children and adolescents One ought to ascertain the number and severity of sore throats particularly in patients suspected of having rheumatic heart disease Nose bleeds may be important because of their frequent occurrence in patients with rheumatic heart disease and pulmonary stenosis

The central nervous system should be considered in regard to the occurrence of "anoxic spells (see p 277) their time of appearance duration frequency and nature It is also important to note specifically

whether they are accompanied by loss of consciousness and by convulsions The symptoms of chorea should be inquired into carefully as well as the mental development of the child The bones joints muscles and skin should be subject to inquiry particularly in regard to the occurrence of rashes petechiae rheumatic nodules growing pains and arthritis The number of joints involved the duration of involvement the presence of swelling heat and tenderness are all important pieces of information

The family history can usually be covered fairly briefly The essential points to remember are the numbers of miscarriages and stillbirths familial occurrence of congenital anomalies and the presence of rheumatic fever or any other type of heart disease Also the question of consanguinity should be raised in every instance

The prenatal history is important particularly in regard to the first trimester Inquiries should be made about bleeding toxemia febrile illness and exposure to communicable diseases Questions concerning the birth and type of delivery should be made Particular emphasis should be placed on the occurrence of cyanosis the use of resuscitative measures and the administration of oxygen all signs of fetal distress Evidence of congenital heart disease is often suggested by a history of delayed discharge from the hospital after birth or of failure to regain birth weight in the expected time

Feeding development immunization and past history require the type of attention customary in the taking of a good pediatric history

PHYSICAL EXAMINATION

The physical examination of the child suspected of having heart disease should follow along well established pediatric lines It is valuable to have a routine sequence which will reduce omissions to a minimum The following points require particular emphasis

General Description

Does the child appear ill or is he happy and playful? Is he in distress of any sort and if so what kind? The body build should be expressed by recording the absolute weight and height in terms of percentiles on a well accepted developmental chart This latter point is particularly important as one follows a patient along through the years and watches his development the influence of intercurrent illnesses and the effects of surgical intervention The phrases "well developed and well nourished" are particularly irritating and meaningless unless they are supplemented by percentiles of the average A developmental chart should be part of the record of every child with heart disease

No general description of a child suspected of having congenital heart disease should fail to mention his preferred posture particularly



Figure 1 Child squatting

the presence or absence of squatting (Fig 1) The color should be noted with particular attention to cyanosis If cyanosis is present its depth and distribution pattern are always pertinent Sometimes patients with minimal arterial unsaturation (such as isolated pulmonary stenosis Ebstein s disease or pulmonary hypertension) are not frankly cyanotic their cheeks and lips have a peculiar high color however with a slight bluish tinge Noticing this the experienced observer will estimate the arterial oxygen saturation to be in the 85 to 90 per cent range Often this type of color is misleading however and even the trained clinician may mistake the flush of a healthy child entering a warm room for borderline cyanosis

Jaundice is less common in children with heart disease than in adults Pallor is obviously an important sign the particular discrepancy of pallor and a relatively adequate hemoglobin level is characteristic of the rheumatic patient The yellowish pallor (*café au lait*) is typical of subacute bacterial endocarditis The particular combination of cyan osis and pallor in patients with cyanotic heart disease and relative anemia is known as "Picasso blue" in our Clinic

Head and Neck

The conjunctivae should be inspected with particular care in regard to the presence of anemia or cyanosis It is surprisingly easy to examine the fundi of most children sufficiently well to note the presence or ab sence of papilledema and venous engorgement Carious teeth are unfortunately common in patients with cyanotic congenital heart disease and may be the source of subacute bacterial endocarditis in these pa

tients as well as in those with rheumatic fever Although the tonsils are not regarded with as much awe today as they were twenty five years ago nevertheless the connections between streptococcal sore throat and rheumatic fever on the one hand and membranous tonsillitis and diphtheritic heart disease on the other are significant enough to warrant careful evaluation of these organs

Inspection of the neck can furnish important clues Vigorous arterial pulsations commonly can be seen in patients with severe aortic disease (coarctation of the aorta patent ductus arteriosus or aortic regurgitation) and in children with ventricular septal defect they are practically unknown in instances of atrial septal defect Distention and pulsation of the jugular veins will be discussed later suffice it to say here that they furnish the best indirect information about the happenings in the right atrium A thrill palpable at the suprasternal notch is practically pathognomonic of one of four conditions aortic stenosis coarctation of the aorta pulmonary stenosis or patent ductus arteriosus Evidence of cervical lymph nodes may be extremely helpful in the diagnosis of rheumatic fever and less commonly of diphtheritic heart disease or even infectious mononucleosis The rare instance of thyrotoxic heart disease may be suspected if goiter is discovered A thrill and continuous murmur localized over the neck should raise the suspicion of an arteriovenous fistula Finally a continuous murmur of evanescent character heard over the neck vessels constitutes a venous hum The differential diagnosis of this will be discussed in some detail in the chapter relating to patent ductus arteriosus (see p 352)

Heart

Inspection and Palpation The examiner should start by noting the presence or absence of deformities in the chest The symmetrical increase in anteroposterior diameter is characteristic of the emphysematous patient Vigorous pulsation of a significantly enlarged right or left ventricle leads to visible left sided chest prominence (congenital heart disease with left to right shunt or rheumatic valvular disease) Right sided chest prominence is present occasionally in patients having tricuspid atresia Visible pulsations sometimes rocking the entire chest are characteristic in certain patients with congenital heart disease involving tremendous shunts (patent ductus arteriosus or large ventricular septal defects with aortic regurgitation) and in children with rheumatic aortic regurgitation The presence of thrills and their timing intensity and localization should be noted The cardiac impulse should be observed with great care [102 362] A good estimation of the hypertrophy of the right or left ventricle can be achieved by noting the point of maximal impulse of the heart beat If this is at the xiphoid process or the lower left sternal border with visible up and down pulsations the chances are that the right ventricle is dominant Conversely if the cardiac impulse is maximal



Figure 2 Comparison of large adult size and small child size stethoscopes with open bell and diaphragm chest piece

at the apex with visible outward pulsations in this area the left ventricle is likely to be enlarged A cardiac impulse palpable at both the xiphoid and the apex strongly suggests combined hypertrophy The character of the impulse may be tapping and quick suggesting a relatively large volume work or heaving and slow indicating increased pressure work Percussion of the entire cardiac silhouette is tiresome time consuming and barely justifiable in this age of easy availability of radiograms At its best this procedure can give the physician only an approximate idea of the size of the heart at its worst it may be completely misleading

Auscultation After inspection and palpation have furnished information regarding the size of the heart and the intensity of its pulsations the examiner should concentrate on auscultation To obtain maximal benefit from this most valuable part of the physical examination the proper instrument must be used The usual "pediatric stethoscope" with the flat chest piece edged with rubber is totally useless for reliable auscultation of the heart [78] Almost equally useless in the examination of infants are the huge adult stethoscopes especially the ones with only a diaphragm chest piece these instruments when placed on the tiny chest of a baby cover practically the entire cardiac silhouette A third kind of stethoscope should be used only when necessary—someone else s stethoscope The pernicious practice of picking up any stethoscope in a hospital ward is to be deplored A stethoscope is a personal instrument much like a fountain pen Best results can be achieved only by the continuous use of the same ear and chest pieces Not everyone s aural canals are of the same size and shape The ear pieces have to fit

the canals tightly without discomfort to assure optimal auscultation[818] It is to be hoped that sartorial considerations will never prohibit the doctor from carrying his own stethoscope in his pocket

What type of stethoscope should be carried for the examination of children with heart disease? It should be a so called binaural stethoscope with a combined bell and diaphragm chest piece The chest piece should be the smallest available the bell 20 mm the diaphragm 30 mm (Fig 2) Even this size bell is sometimes too big for a baby and careful localization of the sounds and murmurs can be achieved only by unscrewing the black Bakelite portion and listening with the small metal end of the chest piece The length of the rubber tubing should probably not be longer than 10 to 12 inches the bore 1/8 inch [81] One ought to listen not only at the apex and the other conventional points but also at selected areas over the entire cardiac silhouette The neck and the chest should be carefully mapped out with the stethoscope using the bell piece first the diaphragm later Low frequency murmurs are best heard with the bell whereas the higher frequencies are best perceived through the Bowles diaphragm If possible the first attempt at listening to any patient s heart should be with the patient lying flat to be followed by turning him on his left side and finally by having him sit up When examining a small infant one cannot be too demanding and one should examine the heart in the position most comfortable to the child irrespective of the gymnastics it may require on the part of the doctor It is remarkable how many children who are completely unmanageable if put on an examining table will be quiet and cooperative when held on the mother s lap

HEART SOUNDS On auscultation the first things to be noted are the rate and the rhythm after which one ought to concentrate on the heart sounds I have found it useful to try to listen to the sounds one at a time aiming to determine the following characteristics of each (1) Where is it heard maximally (apex second left interspace and so forth)? (2) What is its absolute intensity? (3) Where is it observed minimally? (4) What is the degree of splitting? This type of analysis of the first second third and possibly fourth heart sounds should precede the evaluation of murmurs and is of the greatest importance A loud first sound at the apex occurs commonly in mitral stenosis A loud late systolic sound at the pulmonary or aortic area has been observed in aortic or pulmonary stenosis [82] The booming second sound at the pulmonary area is pathognomonic of pulmonary arterial hypertension [84] whereas the absence of this sound at the second left interspace is suggestive of pulmonary stenosis A well defined third heart sound at the apex is common in normal adolescents on the other hand it may be the precursor of a mid diastolic rumble A fourth or atrial sound may be specific for conditions with right atrial hypertension (severe pulmonary stenosis Ebstein s anomaly and total pulmonary venous anom

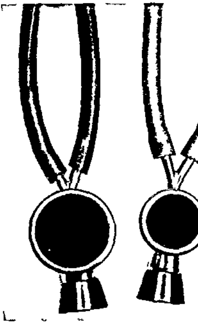


Figure 2 Comparison of large adult size and small child size stethoscopes with open bell and diaphragm chest piece

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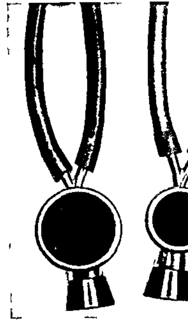


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Abdomen

Gentle often superficial palpation is needed to discover the liver edge in a young infant In normal infants and children the liver may be palpable as far down as 3 cm from the right costal margin in the right nipple line Daily observation regarding the size of the liver is one of the most reliable guides in the estimation of congestive failure in chil dren Frequently one sees a child admitted to the hospital in congestive failure due to paroxysmal atrial tachycardia with a liver estimated to be 7 cm below the right costal margin Adequate treatment of such a patient may result in the absence of a palpable liver within thirty six hours

Although many young children normally have a palpable spleen definite splenomegaly is one of the cornerstones in the diagnosis of sub acute bacterial endocarditis By contrast an enlarged spleen is seldom caused by congestive failure Abdominal situs inversus with or without thoracic situs inversus may suggest associated congenital heart disease The obvious importance of ascites requires diligent search for abdominal fluid Palpation of the abdominal aorta is possible in many children and may be used in the accurate localization of a coarctation of the aorta

Extremities

Cyanosis of the fingers strongly suggests congenital heart disease In patients in whom a definite difference between the coloring of the upper and lower extremities is noted a shunt from the pulmonary artery to the aorta through the ductus arteriosus may strongly be suspected British authors[383] recommend the use of a hot bath to bring out the cyanosis in the lower extremities

Clubbing of the fingers and toes commonly accompanies cyanosis but may be seen in patients without arterial unsaturation (p 276) Clinically clubbing has at least two components one is the rounding of the fingers especially the thumbnails the other is a thickening and shininess of the terminal phalanges with disappearance of the normal creases (Fig 3) In some patients the fingernails show the greatest abnormality in others the changes in the phalanges dominate In children in whom a difference in degree of cyanosis between fingers and toes is noticed the same discrepancy in the degree of clubbing will be observed

Patients with congenital heart disease often have fiery red fingers and toes We have learned to recognize this phenomenon as a precursor of cyanosis and clubbing it is fairly characteristic of those whose arterial oxygen saturation is permanently or paroxysmally low normal or slightly abnormal (±90 per cent) (Ebstein s anomaly isolated pulmonary stenosis with foramen ovale or ventricular septal defect and pulmonary vascular obstruction) The temperature of the extremities should be noted particularly in regard to differences between hands and feet the

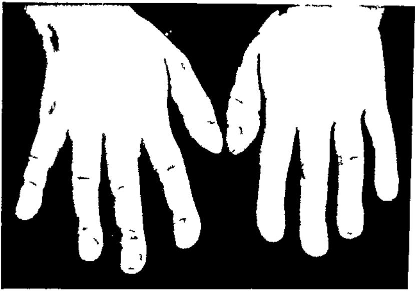


Figure 3 Clubbed fingers

discrepancy may be rather marked in coarctation of the aorta The joints should be inspected carefully heat tenderness redness and swelling are indispensable features in the diagnosis of rheumatic arthritis Rheumatic joints are painful and tender they may easily be differentiated from the nonspecific mild joint pains secondary to orthopedic conditions (flat feet and so forth) Rheumatic nodules (Fig 4) are most commonly found over the knuckles the elbows and the knees Sometimes they can be seen first and felt only later They may be recognized better in indirect light as pea sized nontender structures attached to fasciae or tendon sheets which are freely movable with the skin

Rashes especially erythema marginatum (Fig 5) may be seen in patients with active rheumatic fever Petechiae are characteristic of subacute bacterial endocarditis Deep tender purple spots at the ends of the fingers representing minute emboli (Osler nodes) are practically pathognomonic of subacute bacterial endocarditis Pitting edema of the extremities is not common in children if present it may indicate right sided congestive failure

Palpation of both radial and femoral pulses is of utmost importance Strong radial pulses with weak and late or absent femorals are diagnostic of coarctation of the aorta In this and in congenital aortic stenosis a difference between the strength of the left and right radial pulses is also commonly noted Respiratory variations in pulse volume (pulsus paradoxus) of more than 10 mm of mercury mean pericardial tamponade alternation of only a few millimeters of mercury is indicative of myocardial failure

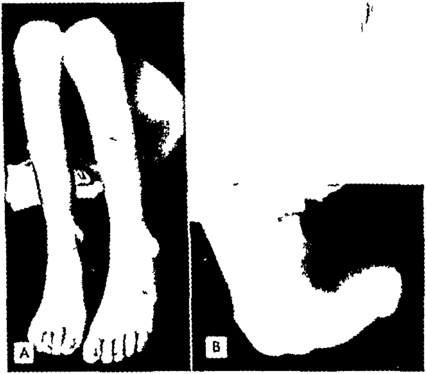


Figure 4 Rheumatic nodules



Figure 5 Erythema marginatum

CIRCULATORY SYSTEM

Arterial Pressure Determination of blood pressure in the arm[10] should be an integral part of every examination It is important to measure the systolic blood pressure in the lower as well as in the upper extremities in all patients suspected of having congenital heart disease thus coarctation of the aorta will not be missed by clinical examination

The auscultatory method using a mercury sphygmomanometer is an easy and well accepted procedure in adults and older children The results can be interpreted without much hesitation

In children especially in the younger age groups the problem is far from simple To begin with the patient usually does not cooperate Second limbs of different sizes require different cuff sizes Finally the results have to be interpreted according to the age of the patient

To ensure optimal cooperation it is worth while if possible to explain the procedure to the child beforehand Often an older child may be distracted by watching the up and down movements of the mercury column when they are pointed out Infants often have to be pacified with a bottle of milk while the blood pressure is being taken Finally in some instances sedation with barbiturates or morphine may be neces sary to ensure some degree of cooperation

The size of the cuff is extremely important in obtaining results comparable to those of arterial blood pressure determined by direct arterial puncture As a rough guide the cuff should cover approximately two thirds of the upper arm or leg A cuff that is too small gives falsely high readings For accurate results the pediatrician should have available at least the following cuff sizes 3 5, 12 and 18 cm widths Sometimes a larger cuff folded over may be adapted to a smaller limb The bag should be completely deflated and applied evenly snugly without bulg ing around the upper arm or leg and should completely encircle the limb

Once the cuff of proper size has been applied to the patient it may be well to obtain a quick reading of the systolic pressure by palpation The cuff should be inflated to a point well above the disappearance of the radial pulse Then gradually (2 to 3 mm of mercury per beat) the pressure should be released until the radial pulse reappears This point clearly corresponds to the systolic pressure determined by direct meas urement

Once the systolic pressure has been determined by palpation the systolic and diastolic pressures should be estimated by the usual aus cultatory method According to the latest report of a committee of the American Heart Association the diastolic pressure corresponds to the level at which the auscultatory sounds completely disappear[11] Simul taneous measurements of the diastolic pressure by the sphygmoman ometer and by an indwelling arterial cannula reveal this not to be the case in every instance Therefore if the sounds become obviously muffled

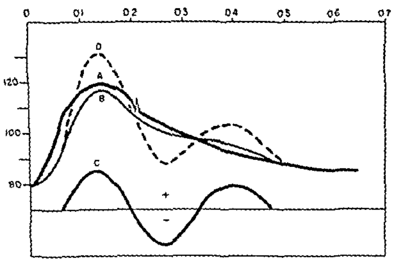


Figure 6 Diagram illustrating the principles of damping and wave summation in the arterial system (*a*) Aortic pressure pulse (*b*) Changes in contour and amplitude due to damping (*c*) Nature of rebound wave (*d*) Summation of curves *b* and *c* (ordinate pressure in mm Hg abscissa line in 0 1 second) (From C J Wiggers Modern Medical Monographs New York Grune & Stratton Inc)

at one pressure level and disappear completely at a lower one both figures should be recorded since either may represent the true diastolic pressure

Though the auscultatory method may not be feasible in a restless infant and the palpatory method may not be adaptable in patients who do not have easily palpated pulses in the lower extremities (coarctation of the aorta) the so called flush method[1, 6] can be used in both situations A cuff of appropriate size is placed around the ankle or wrist The limb is elevated and massaged to produce blanching The cuff is rapidly inflated to well above the expected systolic level Then the limb is lowered to approximate heart level and the cuff is slowly gradually deflated The point at which a marked flush appears below the cuff in the previously blanched limb is taken to correspond to the systolic pressure The flush pressure is usually lower than the systolic peak pressure obtained by direct arterial puncture or by the auscultatory method It commonly approximates the level of the mean arterial pressure With these limitations the flush pressure is an extremely useful method of obtaining information about the blood pressure of certain infants when no other method is applicable Perhaps its prime usefulness is in revealing relative hypertension of the arms and thus proving the presence of coarctation of the aorta in instances in which this diagnosis could not be made by any other method

The systolic pressure in the lower extremities is usually significantly higher than in the upper extremities because of a summation effect of

the forward pulse wave with the rebound wave (Fig 6) A reversal of this relationship is pathologic and implies the presence of an aortic block The diastolic pressures in the upper and lower extremities should be identical a significant difference usually means that the cuff over the leg was relatively small

The actual height of the systolic pressure has to be interpreted in relation to the patient s age Various authors have placed the systolic blood pressure level in normal infants at approximately 70 to 90 mm of mercury between birth and one year of age Premature infants have correspondingly lower pressures Between the ages of one and five years the average systolic pressure rises to about 100 mm of mercury between five and fifteen years it gradually climbs to 120 mm of mercury The diastolic pressure during the same period changes relatively little from 55 to 61 mm Average blood pressures in infants and children taken from the most reliable sources in the literature are given in Table I

Table I Normal Blood Pressure for Various Ages

(Adapted from data in the literature)
(Figures have been rounded off to nearest decimal place)

Ages	*Mean Systolic* ± 2 S D	*Mean Diastolic* ± 2 S D
Newborn	80 ± 16	46 ± 16
6 mos –1 year	89 ± 29	60 ± 10†
1 year	96 ± 30	66 ± 25†
2 years	99 ± 25	64 ± 25†
3 years	100 ± 25	67 ± 23†
4 years	99 ± 20	65 ± 20†
5–6 years	94 ± 14	55 ± 9
6–7 years	100 ± 15	56 ± 8
7–8 years	102 ± 15	56 ± 8
8–9 years	105 ± 16	57 ± 9
9–10 years	107 ± 16	57 ± 9
10–11 years	111 ± 17	58 ± 10
11–12 years	113 ± 18	59 ± 10
12–13 years	115 ± 19	59 ± 10
13–14 years	118 ± 19	60 ± 10

* From R J Haggerty M Maroney and A S Nadas Essential Hypertension in Infancy and Childhood Am J Dis Child to be published [37]

† In this tudy the point of muffling was taken as the diastolic pressure

The most common causes of persistent and significant elevation of the systolic blood pressure in children listed in the approximate order of their frequency are (1) renal disease (2) coarctation of the aorta (3) widened pulse pressure (aortic regurgitation patent ductus arteriosus thyrotoxicosis) (4) diseases of the central nervous system (polio myelitis tumors encephalitis) (5) poisoning with mercury (acrodynia) and lead (6) endocrine disorders (pituitary adrenal tumors pheo chromocytomas) and (7) essential hypertension

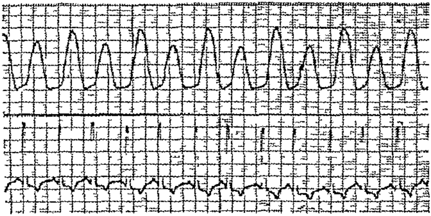


Figure 7 Pulsus alternans in patient with severe aortic stenosis Note on top line the alternating height of systolic pressure within the left ventricle and on the bottom the electrocardiogram with QRS complexes of approximately identical height

Low blood pressure is most commonly explained by the presence of shock or so called vasovagal syncope (fainting)

Pulse Pressure The pulse pressure may be calculated by subtracting the diastolic from the systolic pressure A widened pulse pressure detectable by the bounding water hammer type of radial pulse is commonly seen (1) in fever anemia thyrotoxicosis and after vigorous exercise (2) in aortic regurgitation patent ductus arteriosus and arteriovenous fistulas and (3) in complete heart block

Conversely narrow pulse pressures may be observed in (1) shock (2) severe mitral or aortic stenosis (3) pericardial tamponade or constrictive pericarditis and (4) certain instances of congestive failure

Two peculiarities of the pulse may best be detected while determining the blood pressure (1) Pulsus paradoxus characteristic of the accumulation of an appreciable amount of fluid in the pericardial sac consists in a marked decrease of systolic pressure and narrowing of the pulse pressure on deep inspiration (2) Pulsus alternans a sign of left ventricular failure consists in an appreciable drop in systolic pressure in alternate beats (Fig 7)

Venous Pressure Determination of venous blood pressure and analysis of the jugular venous pulse waves are extremely useful tools in cardiovascular diagnosis but are not nearly as widely used as is the determination of the arterial blood pressure The reason for this neglect may well be the absence of a simple accurate apparatus not involving venipuncture similar to the sphygmomanometer

In spite of the technical difficulties a great deal of attention has been paid to the jugular venous pulse in adults by careful cardiologists The accurate determination of the venous pressure and detailed analysis of the venous pulse waves are well nigh impossible to obtain in infants because of their lack of cooperation

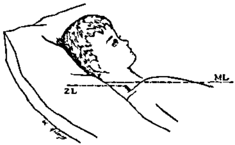


Figure 8 Patient lying down at approximately 45 degrees the pressure (*ZL*) in the right atrium does not rise above the manubrium line (*ML*) (After a drawing from T Lewis Diseases of the Heart Described for Practitioners and Students New York Macmillan Company)

In children who cooperate especially the older ones simple inspection furnishes reliable information regarding venous pressure and thus indirectly the right atrial pressure Perhaps the simplest and most reliable test is inspection of the jugular vein of the patient when he is erect According to Lewis [09] the external jugular vein should not be visible above the suprasternal notch unless the venous pressure is elevated A crude estimation of venous pressure is furnished by measuring the height of the jugular venous column vertically in a standing patient above the suprasternal notch If the patient is recumbent the height of the jugular venous column should not rise above an imaginary straight line across the manubrium of the sternum (Fig 8)

These rough clinical guides usually give satisfactory answers only in regard to the presence or absence of gross pressure elevation For purposes of accurate determination however direct measurements by means of venipuncture are necessary Recently an apparatus for determining venous pressure in this way has been made available commercially [57] Properly used this gives accurate and reproducible results

A simple apparatus which may be made available in the treatment room of any hospital consists of a spinal manometer connected to a T tube and mounted on a stand The zero level of the manometer should be adjustable One end of the T tube is connected to a length of rubber tubing with a glass adapter to which a number 20 to 22 needle is attached The other end of the T tube is connected with a reservoir by rubber tubing and should have a clamp on it The reservoir contains saline solution with a drop of heparin

The patient is propped up at an approximately 45 degree angle with his arm resting on a pillow All clothing is loosened and the patient should be as relaxed as possible The zero level of the manometer is adjusted to the level of the manubrium of the sternum If the test is performed with the patient lying flat on an examining table the zero level should be adjusted at 5 5 cm above the table top It is extremely important to record the point at which the zero level of the manometer was placed only in this way can the results have any comparative value The manometer is filled with saline solution from the reservoir and the rubber tubing is clamped An antecubital vein is entered and the saline column in the meniscus of the manometer then indicates the height of

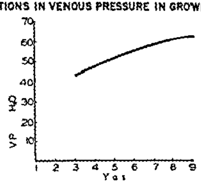


Figure 9 Venous pressure in children (From G E Burch A Primer of Venous Pressure Philadelphia Lea & Febiger)

the venous pressure in millimeters of saline It is important to watch the rise and fall of the saline column with respirations for a few minutes before a definite reading is taken to see whether it is in free equilibrium with the venous system The height of the mean venous pressure in children between the ages of three and five years has been established by Burch[57] as being 40 mm of water and between the ages of five and ten years 58 mm of water (Fig 9)

Not only is the height of the venous pressure of great importance in the clinical diagnosis of heart disease but the pattern of the jugular venous pulsation is of considerable interest as well By careful inspection in a patient with superficial jugular veins three separate waves may be noted throughout every cardiac cycle For accurate analyses of these waves the jugular venous pulse should be registered The instrument most commonly used today for this purpose is a two channel "twin beam photographic machine One of the galvanometers registers the venous pulsations the other the electrocardiogram for a reference tracing Further elucidation of the jugular venous pattern may be achieved by using a phonocardiogram as a reference tracing (Fig 95 p 96)

If the jugular venous tracing thus registered is analyzed three separate waves can be distinguished (Fig 10) (1) The a wave or presystolic wave immediately follows the p wave and precedes the q wave on the electrocardiogram By physical examination it can be recognized as the wave of the jugular venous pulse which immediately precedes the apical impulse of the heart It originates from the reflux in the venae cavae caused by atrial contraction (2) The c wave or early systolic wave is synchronous with the plateau of the carotid pulse hence the designation c It usually follows one or two seconds beyond the termination of the QRS complex in the electrocardiogram It is said to originate partly from the isometrically contracting right ventricle pushing back against the closed tricuspid ring and partly from the impulse of the carotid artery itself (3) The v wave marks the end of ventricular

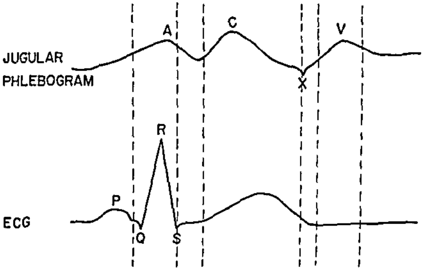


Figure 10 Jugular phlebogram Schematic drawing indicating A C and V waves

systole It occurs at the time of or slightly after the dicrotic notch at the carotid tracing usually some distance beyond the T wave of the electro cardiogram and just beyond the end of the second sound in the phono cardiogram This v wave is caused by the gradual filling of the right atrium with blood from the systemic venous return

Analysis of these elements of the jugular venous pulse reveals a certain characteristic pattern A large a wave indicating powerful atrial systole can be seen characteristically in the phlebogram of conditions causing marked "P pulmonale" such as pulmonary stenosis with in tact ventricular septum tricuspid stenosis or atresia and Ebstein s ano maly Obviously no a waves may be seen in atrial fibrillation A waves not followed by other venous pulse waves are noted in instances of com plete atrioventricular block An exaggerated v wave is seen in tricuspid regurgitation Tall v waves preceded by systolic collapse have been found commonly in patients with large atrial septal defects

Circulation Time Determination of the circulation time requires only brief mention In pediatric cardiology this measurement is used principally for two purposes (1) The progress of patients in congestive failure may be estimated by repeated determinations of the circulation time This is based on the principle that congestive failure is accom panied in most instances by slow circulation time which improves with treatment By the same token determination of the circulation time auto matically separates the so-called high output failures with rapid circula tion time (thyrotoxicosis anemia or beriberi) from the conventional cases of congestion with slow circulation time (2) A significantly shortened circulation time in patients with congenital heart disease indicates a right to left shunt

Methods for the determination of circulation time are manifold In

Table 2 Circulation Time by Fluorescein Method in Normal Infants and Children

Author	Distance	Number of Patients	Age	Average and Range in Seconds
Lexer et al [1]	Hand to conjunctiva	55	0–5 days	10 2 (6–14)
Wirzberger et al [2]	Arm to lip	25	1–24 months	7 0 (5–9 0)
Wirzberger et al [2]	Arm to lip	51	3–13 years	11 5 (8–16)
Gasul et al [3]	Arm to lip	14	4–24 months	6 5 (5–8 5)
Gasul et al [3]	Arm to lip	36	3–13 years	8 5 (5–12 5)

[1] Am J Dis Child 83 645 1952
[2] J Pediat 22 726 1943
[3] J Pediat 34 460 1949

Table 3 Arm-to-Tongue Circulation Time by the Decholin Method in Normal Children (Lind)

Number of Patients	Age in Years	Average and Range in Seconds
24	3–6	10 5 (8–17)
65	6–9	10 4 (7 5–15 5)
67	9–12	11 0 (7 5–14)
76	12–15	11 9 (10–16)

* Acta paediat Scandinav 38 423 1949

The Childrens Hospital in Boston two methods are used principally Both give reliable and reproducible results both measure the arm to mouth circulation time Tables 2 and 3 contain normal values for these two methods

In the fluorescein method 0 07 cc per pound of body weight of a solution containing 5 per cent sodium fluorescein and 5 per cent sodium bicarbonate is injected into the antecubital vein in a dark room A fluorescent light (a modification of the Woods lamp) shines on the patients lips The end point is usually sharp when yellowish fluorescein shows up on the patients mouth The child should be carefully examined under a Woods lamp before the test for fluorescent material which may confuse the reading penicillin and Aureomycin have given trouble in this respect This test is especially useful with small children when an objective end point is needed Doses of more than 5 cc may cause vomiting

In the second method Decholin is used in dosages of 2 cc for children less than seven years of age and 3 cc for older children in order to determine the arm to mouth circulation time The material is injected into an antecubital vein but the end point here is subjective The patient should be instructed to indicate immediately by raising his hand when a bitter taste touches his tongue This test can be used effectively in patients seven years old or older

There is a third method with which I have had no personal experience It consists in the use of Evans blue for the determination of cir

culation time with an ear oximeter registering the end point According to Swan and Wood[3 7] it is a most effective test

ROUTINE LABORATORY TESTS

Routine laboratory tests performed on all children including those with cardiac disease will not be described here in detail Their particular applicability to children with heart disease will however be mentioned briefly

URINALYSIS

Patients with congestive failure may have albuminuria a urine of high specific gravity and mild hematuria Hematuria is also found in patients with subacute bacterial endocarditis

COMPLETE BLOOD CELL COUNT

Leukocytosis of moderate degree may be present in congestive failure subacute bacterial endocarditis and acute rheumatic fever with or without failure A high hematocrit value is always found in patients with cyanotic congenital heart disease In addition to determining the hematocrit level a careful analysis of the blood indices (mean corpuscular volume mean corpuscular hemoglobin and mean corpuscular hemoglobin concentration) as well as inspection of the smear is indicated Such a careful survey of the hematologic data often reveals the presence of hypochromic anemia requiring iron treatment or a dangerously high hematocrit value (more than 80 per cent) requiring venesection The normochromic or hypochromic anemia noted with subacute bacterial endocarditis and rheumatic fever should be observed

ACUTE PHASE REACTANTS

In acute infections particularly rheumatic fever certain substances which wax and wane with the progress or subsidence of the disease appear in the blood These substances are called acute phase reactants Two tests—one used widely the other only sporadically—giving information about these substances should be mentioned

The erythrocyte sedimentation rate is used universally as a reliable index of rheumatic activity An elevated erythrocyte sedimentation rate (Westergren more than 20 mm per hour Wintrobe corrected more than 15 mm) is almost invariably present in patients with acute rheumatic fever One notable exception is the finding of a normal sedimentation rate in some patients with acute rheumatic fever in congestive failure High sedimentation rate levels are also found in patients having subacute bacterial endocarditis The phase reactants contributing to the sedimentation rate are probably fibrinogen alpha and gamma globulin

The C reactive protein is an acute phase reactant not normally present in the blood Chemically the substance is an alpha globulin precipi

tated with the somatic C polysaccharide of pneumococci In the routine laboratory test rabbit serum is used to precipitate the substance from the patient's serum It is present with high regularity in almost all patients who have acute rheumatic fever including those with congestive failure However patients with certain isolated rheumatic manifestations usually those not accompanied by fever or elevated sedimentation rate (such as chorea erythema marginatum or Aschoff bodies in the left atrium) fail on occasion to show C reactive protein in their blood The principal value of the C reactive protein determination is that it furnishes the best laboratory evidence for subsidence of rheumatic activity Usually the substance disappears from the blood at an appreciable interval after the return of the erythrocyte sedimentation rate to normal

Bacteriologic and Serologic Examination

Direct or indirect evidences of a streptococcal infection are found in the vast majority of patients with acute rheumatic fever Direct proof is the finding of group A hemolytic streptococci in nose and throat cultures from patients having acute rheumatic fever Indirect evidence of a poststreptococcal state is furnished by an elevated antistreptolysin O titer (250 units or more) in the serum A detectable increase in the antistreptolysin titer starts about two weeks after the onset of streptococcal infection The titer usually reaches its maximal level four to six weeks after the onset and returns to normal within four months Although obviously the diagnosis of rheumatic fever cannot be based on the elevated antistreptolysin titer alone its absence excludes for practical purposes the presence of active rheumatic fever

A positive blood culture is an integral though not indispensable part of the diagnosis of subacute bacterial endocarditis

CHAPTER 2

Radiology of the Heart

No cardiac examination is complete without an adequate radiologic survey of the patient A complete radiologic examination should include both radiography and fluoroscopy preferably with appropriate spot films

FLUOROSCOPY

Cardiac fluoroscopy is the direct examination of the patient s chest and heart under a fluoroscopic screen Although the proximity of the screen to the chest inevitably results in enlargement and distortion of the cardiac silhouette fluoroscopy provides certain information not otherwise obtainable

A word should be included here concerning the controversy as to *whether the radiologist or the cardiologist should perform this relatively* simple examination Ideally cardiac fluoroscopy should be a cooperative venture The broad experience of a good radiologist his ability to survey adequately the entire chest and abdomen under the screen to recognize the imprint of vascular anomalies on the barium filled esophagus to note the adequacy of the diaphragms all are invaluable in obtaining a good total picture of the patient On the other hand the familiarity of the cardiologist with the cardiac silhouette and his mental image of the clinical picture and the electrocardiogram of the patient are indispensable in the darkness of the fluoroscopy room Optimally both the radiologist and the cardiologist should be present at fluoroscopy If however there has to be a choice between the two I believe that unless the radiologist has made a special study of children with heart disease the pediatric cardiologist is probably better equipped to perform the fluoroscopic examination

The most obvious advantage of cardiac fluoroscopy over any other type of roentgenologic technique is that it presents the heart in a dynamic fashion one may thus obtain a good image of the amplitude of the heart beat The second advantage of this method is that by rotation of the patient to the oblique and lateral positions the examiner may bring hidden structures into view on the fluoroscopy screen Third such extracardiac structures as the thymus can be separated by fluoroscopy from the cardiac silhouette proper Fourth by having the patient swallow a barium mixture one can easily outline the esophagus and thus demonstrate the imprints of adjoining portions of the heart and great vessels

Last but not least cardiac fluoroscopy at a glance gives a great deal of information about the other intrathoracic organs

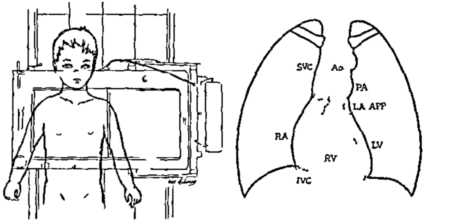
The fluoroscope to be used in pediatric cardiology should be equipped with a tilt table allowing for the examination of infants lying down and older children standing up A device for taking spot films is also an important part of the fluoroscope as is a timing device to remind the examiner of the amount of exposure the patient has had No more than five to ten minutes of fluoroscopy time at 50 to 70 kilovolts should be used for any one patient

One cannot emphasize strongly enough the necessity for a dark adaptation period of at least five minutes behind red glasses before starting fluoroscopy This time can be spent usefully in explaining to the parents and the child what is going to happen In this way the whole procedure is a great deal less frightening Also it gives the child a chance to look around the room and see the people in it Another point worth emphasizing is that it is almost impossible to use the fluoroscope on a child especially a small one without the help of a trained nurse or a technician her assistance is needed to hold the child and to offer the barium meal in a persuasive fashion

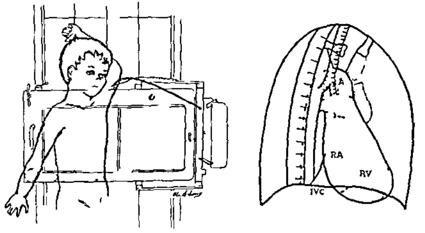
If the child is cooperative and understands what is required of him fluoroscopy in the upright position is preferable Otherwise much better results can be obtained if he lies on his back with his arms and legs restrained The first view under the fluoroscope should be of the entire chest This gives the examiner an opportunity to see the heart size in relation to the entire thoracic cavity and to notice the rate of respiration the movement of the diaphragms and the site of the stomach bubble After this general orientation the screen should be narrowed down to the cardiac silhouette and both hili The child should then be turned from the posteroanterior view to the two obliques (Fig 11) He should be examined in at least these three views and preferably in many others Certain structures such as the main pulmonary artery often are seen best somewhere midway between the posteroanterior and the right anterior oblique views

The individual chambers making up the cardiac silhouette in the different views are schematically presented in Figure 11

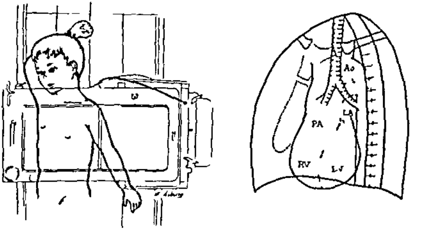
In the posteroanterior view one should try to identify three permanent components of the left border (the left ventricle the *pulmonary* artery and the aorta) and search for the outlines of the left atrium The pulmonary artery may be distinguished from the left ventricle by looking for the rocking beat caused by the contracting left ventricle on the one hand and the expanding main pulmonary artery on the other Identification of the main pulmonary artery segment from the aortic knob should present no major difficulties there is usually a definite indentation between the two structures and the pulsation of the aorta is as a rule considerably stronger than that of the pulmonary artery The aortic knob



A



B



C

Figure 11 Schematic representation of (*A*) posteroanterior (*B*) right anterior oblique and (*C*) left anterior oblique views of the heart at cardiac fluoroscopy

can also be identified by following the continuous sweep of the aorta from the upper right border of the heart up through the sternum where it juts out on the left as the knob If the left atrium projects out on the left border it is represented as a double contour with relatively weak pulsations overlapping the powerful contractions of the left ventricle and the main pulmonary artery If the left atrium is large enough to project out at the left border it usually pushes the barium filled esophagus to the right in the posteroanterior view and posteriorly in the right anterior oblique position

The right border of the posteroanterior view as seen in Figure 11 is normally made up principally of the right atrium and the superior vena cava with the ascending aorta contributing to the upper right border A large left atrium may also participate in the formation of the right border forming a double contour In this case the other evidences of left atrial enlargement will also be noted in the posteroanterior and the right anterior oblique views

The third observation—an extremely important one—to be made in the posteroanterior view is the size of the hilar vessels These are best seen with the fluoroscope concentrated on the vessels ½ to 1 inch away from the cardiac silhouette Both the left and right sides should be surveyed and the size of the vessels and the presence or absence of pulsations should be noted Definite differentiation of intrinsic pulsations of the hilar vessels ("hilar dance") from activities transmitted from the main cardiac silhouette requires considerable patience and experience Two points help in distinguishing these two kinds of pulsations the true intrinsic pulsation is (*a*) concentric and (*b*) present at some distance from the cardiac border

The final observation to be made in the posteroanterior view is the contribution of the thymus to the cardiac silhouette The thymic shadow may make the heart appear much larger than it actually is but it can be separated from the heart relatively easily by noting its "apron like" contour and observing its lower tip running up and down with respirations This distinction is especially necessary in the examination of well nourished infants in whom the thymus is usually large In older children or emaciated infants the problem should not arise

The schematic presentation of the two oblique views is found in Figure 11 The left anterior oblique (LAO) is particularly useful in determining the relative size of the two ventricles Incipient left atrial enlargement may be suspected from the elevation of the left main bronchus in this view The size of the ascending aortic arch as well as that of the right main pulmonary artery may be estimated in this view The right anterior oblique (RAO) view furnishes a useful measure of right ventricular enlargement by demonstrating how much of the anterior border is in direct contact with the sternum The main advantage



Figure 12 Generalized enlargement of the heart in patient with primary myocardial disease Note also the marked pulmonary vascular engorgement

of this view however is in helping to determine even minimal degrees of atrial (especially left atrial) enlargement

Diagnostic Patterns and Their Significance

When cardiac fluoroscopy is carefully performed certain radiologic patterns emerge A brief discussion of these will follow

The cardiac beat may be overactive in anemia thyrotoxicosis large left to right shunts aortic regurgitation and excitement An unusually quiet beat is noted in conditions such as congestive failure myocardial disease and pericardial effusion or constriction Bradycardia of 50 beats per minute or less usually indicates complete heart block and is well recognizable on the fluoroscopic screen by the rapid atrial and slow ventricular pulsations

The general contour of the cardiac silhouette may be typical of certain well defined entities The globular heart involving enlargement of all chambers is characteristic of the primary myocardial disease group (Fig 12) The "water bottle" shape of a large pericardial effusion is easily recognizable (Fig 13) The "sheep's nose" contour or *coeur en sabot* (Fig 14) is typical of the tetralogy of Fallot or of some tricuspid atresias The "figure 8" shape of the heart is seen in total anomalous pulmonary venous drainage into the superior vena cava (Fig 15)

Minimal enlargement of the left atrium can most easily be detected in the right anterior oblique view by the impression it leaves on the barium filled esophagus (Fig 16 A) More severe degrees of left atrial



Figure 13 Posteroanterior view of a heart with pericardial effusion

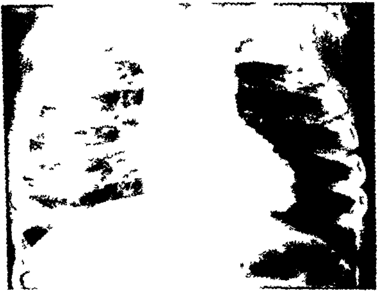


Figure 14 Sheep's nose contour (*coeur en sabot*) of heart of patient with tetralogy of Fallot

enlargement are characterized in the posteroanterior view by the deviation of the barium filled esophagus to the right (Fig 16 B) and in the left anterior oblique view by the elevation of the left main bronchus Left atrial enlargement is seen in cases of left ventricular failure mitral valvular disease patent ductus arteriosus and occasionally in ventricular

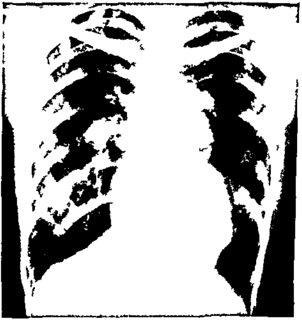


Figure 15 Posteroanterior view of characteristic figure of 8 configuration of heart of patient with total anomalous pulmonary venous drainage into the superior vena cava

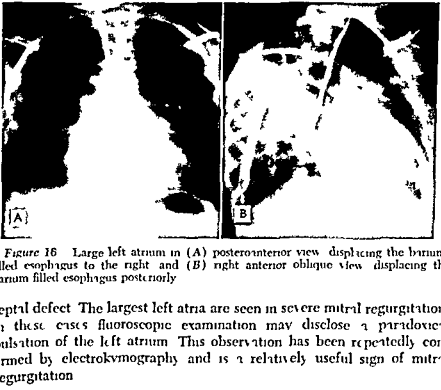


Figure 16 Large left atrium in (*A*) posteroanterior view displacing the barium filled esophagus to the right and (*B*) right anterior oblique view displacing the barium filled esophagus posteriorly

septal defect The largest left atria are seen in severe mitral regurgitation in these cases fluoroscopic examination may disclose a paradoxical pulsation of the left atrium This observation has been repeatedly confirmed by electrokymography and is a relatively useful sign of mitral regurgitation



Figure 17 Left ventricular enlargement in (A) posteroanterior view showing the cardiac apex pointing downward and (B) left anterior oblique view showing the cardiac apex pointing downward and posteriorly



Figure 18 Large ascending aorta in (A) posteroanterior and (B) left anterior oblique views

Enlargement of the left ventricle is best visualized in the postero anterior and the left anterior oblique views (Fig 17) In both of these an enlarged left ventricle manifests itself by producing displacement of the cardiac apex downward and outward In the posteroanterior view the enlarged left ventricle sometimes causes a rounding of the left ventricular segment Left ventricular enlargement is characteristic of mitral regurgi tation aortic valvular disease coarctation of the aorta patent ductus arteriosus certain instances of ventricular septal defect and tricuspid atresia

The aorta can best be outlined in the posteroanterior and the left anterior oblique views (Fig 18) Enlargement of the aortic arch is characteristic of congenital valvular aortic stenosis and aortic regurgita tion Hypoplasia of the aorta is generally said to be typical of atrial septal defect (Fig 19) Vigorous pulsation and collapsing beat of the

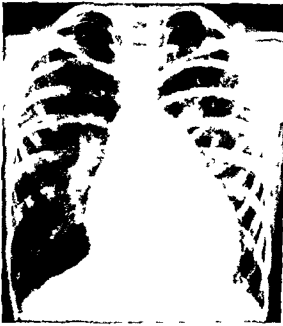


Figure 19 Hypoplastic aorta in posteroanterior view



Figure 20 A Normal left aortic arch with depression on the barium filled esophagus
B Right aortic arch with impression on the esophagus on the right side

aorta are seen in aortic regurgitation patent ductus arteriosus and certain instances of high ventricular septal defect The position of the aortic arch can best be demonstrated by asking the patient to swallow a mouthful of barium paste and watching the passage of this radiopaque material down the esophagus A normal left aortic arch causes slight deviation of the esophagus to the right in the posteroanterior view (Fig 20 *A*) and posteriorly in the left anterior oblique view Conversely if the aortic arch is on the right the esophagus deviates to the left in the posteroanterior view (Fig 20 *B*) and anteriorly in the left anterior oblique position Right aortic arch is commonly seen without any other



Figure 21 Large right atrium in (*A*) posteroanterior and (*B*) right anterior oblique views



Figure 22 Large right ventricle in (*A*) posteranterior (*B*) right anterior oblique and (*C*) left anterior oblique views Note in *B* how the heart displaces the retrosternal space

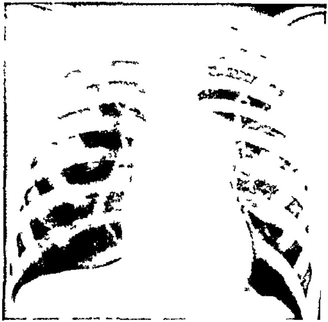


Figure 23 Prominent main pulmonary artery in posteroanterior view

cardiovascular anomaly It is a frequent concomitant of ventricular septal defect with pulmonary stenosis but is practically never seen in conjunction with isolated pulmonary stenosis The diagnosis of so called vascular rings by means of a barium swallow is reviewed in some detail on page *415 ff*

The superior vena cava forms the upper right border of the cardiac silhouette in the posteroanterior view Tremendous dilatation of this structure may be seen in cases of total anomaly of the pulmonary venous drainage Enlargement to a lesser degree is a fairly constant concomitant of the tetralogy of Fallot syndrome

Right atrial enlargement can best be demonstrated in the posteroanterior view and in the right anterior oblique view (Fig 21) An enlarged right atrium is seen with tricuspid disease Ebstein s anomaly pulmonary stenosis pulmonary vascular obstruction and atrial septal defect The largest right atria are seen in tricuspid regurgitation and Ebstein s disease

Right ventricular enlargement may best be visualized in the posteroanterior view by an upward and outward displacement of the cardiac apex (Fig 22 *A*) If the right ventricle is large it may totally displace *the retrosternal space in the right anterior oblique view (Fig 22 B)* In the left anterior oblique view the generally rounded contour with an anterior bulge is characteristic of an enlarged right ventricle (Fig 22 *C*) Right ventricular enlargement is always present with tricuspid regurgitation pulmonary stenosis pulmonary vascular obstruction mitral stenosis large ventricular septal defect and atrial septal defect

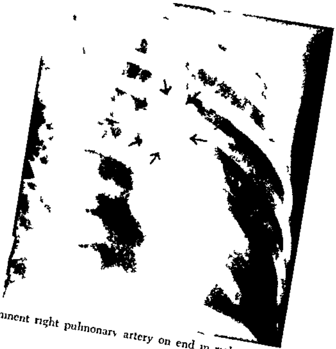


Figure 24 Prominent right pulmonary artery on end in right anterior oblique vie

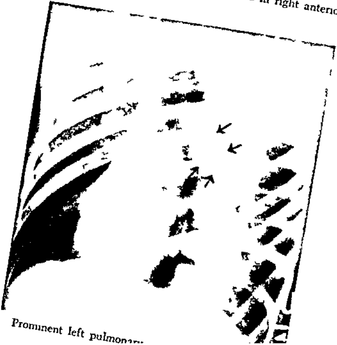


Figure 25 Prominent left pulmonar

The main pulmonary artery segment is best outlined in the postero anterior view or with the patient turned a few degrees from the posteroanterior toward the right anterior oblique (Fig 23) The right main pulmonary artery is best seen on end in the right anterior oblique position just below and posterior to the ascending aorta (Fig 24) The left main pulmonary artery is clearly outlined in the left anterior oblique position (Fig 25) below and to the left of the ascending aorta The pulmonary arteries may be enlarged in pulmonary hypertension valvular pulmonary stenosis and patent ductus arteriosus Hypoplastic pulmonary artery or absence of the main pulmonary artery segment is typical of infundibular pulmonary stenosis and transposition of the great vessels Hyperactive pulsations in the main pulmonary artery segment are characteristic of large left to right shunts pulmonary vascular obstruction and certain cases of mitral valvular disease In patent ductus arteriosus the amplitude of pulsations in the aorta and the pulmonary artery is frequently identical in intensity and synchronous in timing

The status of the pulmonary vasculature can best be judged by radiography Cardiac fluoroscopy gives invaluable information however about the presence or absence of intrinsic pulsations so called hilar dance This is noted in all instances of congenital heart disease with a large left to right shunt and in cases of pulmonary hypertension Expansile pulsations are almost never seen with acquired heart disease even in the presence of marked pulmonary vascular engorgement

RADIOGRAPHY

The patient s chest should be in direct contact with the film and the tube should be 6 feet from the patient In this fashion distortion of the cardiac silhouette is kept at a minimum (10 per cent) and the image is reproducible As a rule three views are taken anteroposterior left anterior oblique and right anterior oblique with left or right laterals

The advantages of radiography over fluoroscopy are that a much higher degree of definition can be achieved permitting greater recognition of detail and of course the fact that a permanent record is obtained This record is invaluable in the long term study of any single patient or disease

Cardiac Silhouette

The contour of the normal heart as seen in the posteroanterior radiogram depends somewhat on the height of the diaphragm For this reason if possible all films should be taken in either full inspiration or full expiration (Fig 26)

The age of the patient is also an important consideration in judging the cardiac silhouette The heart of a normal newborn or young infant has a right ventricular contour corresponding to the dominance of the right ventricle in this age group This results in a *coeur en sabot* type

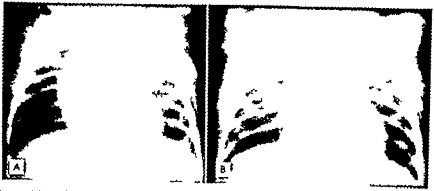


Figure 26 Change in heart size caused by respirations (*A*) Full inspiration (*B*) Expiration

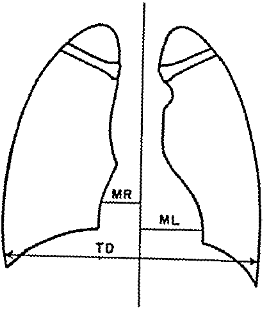


Figure 27 Diagram for determination of cardiothoracic ratio

of appearance The pathologic types of right ventricular enlargement such as tetralogy of Fallot can be seen only as an exaggeration of the normal configuration From the age of approximately six months on the heart starts assuming the "normal left ventricular configuration This process reaches its culmination in the older age group in which the patient with hypertensive disease has definite left ventricular enlargement The left ventricular type of configuration in a newborn or young infant is as distinctly abnormal as is a right ventricular shape in an older child

Innumerable tables for the determination of the "normal heart size"

for any age height or weight are available They are cumbersome difficult to use and far from accurate for children For practical purposes the use of the cardiothoracic ratio has proved to be as satisfactory as any method and it is by far the simplest The ratio consists of the relation of the transverse diameter of the heart to the maximal internal width of the chest (Fig 27) The transverse diameter is the sum of the midleft and midright diameters these are determined by drawing a horizontal line from the midline to the farthest left and farthest right borders of the heart A cardiothoracic ratio of 0 5 that is a heart which is less than half of the size of the thorax usually means a heart of average size This ratio cannot be used with any accuracy in newborn and small infants here a transverse diameter of 5 5 cm is taken as the upper limit of normal All these total measurements are approximations at best The size of the individual chambers may be more valid than the determination of the size of the heart as a whole However changes in the cardiothoracic ratio through the years in one patient may well reflect the *progress of heart disease*

Pulmonary Vasculature

The lung fields may be divided arbitrarily into three vertical sectors the hilus the middle and the outer third Normally the vascular markings are easily observed in both hili (Fig 28 *A*) Because of the position of the cardiac silhouette the right hilus is better visualized in the anteroposterior view than in the left The vascular elements of the hilar markings consist of vessels seen on end plus the left and right main pulmonary arteries seen longitudinally Normally vessels of appreciable caliber are seldom seen in the middle third of the lung field and never *in the outer third Presence of such vessels indicates pulmonary vascular* engorgement (Fig 28 *B*)

One may assume that the engorgement is active if the enlarged vessels have clear sharply defined edges Haziness poor outline and general "filminess" of the lung field are characteristic of passive congestion of pulmonary edema The differential diagnosis between passive congestion and pulmonary infection is sometimes extremely difficult even impossible as a rule however passive congestion is best seen in *the hilus and the middle third of the lung field* and is uniformly distributed over both sides as well as over the apex and the base Infectious processes in contrast are usually more spotty in distribution and may extend throughout the outer third of the lung field without seriously affecting the hilus A determination of the peripheral pulmonary vasculature also may best be judged by radiography If the vessels—on end or lengthwise—are small in the middle or outer lung field we obtain the picture of pulmonary ischemia (Fig 28 C) This in fact is the result of normal or diminished amount of blood with low pressure in the lungs

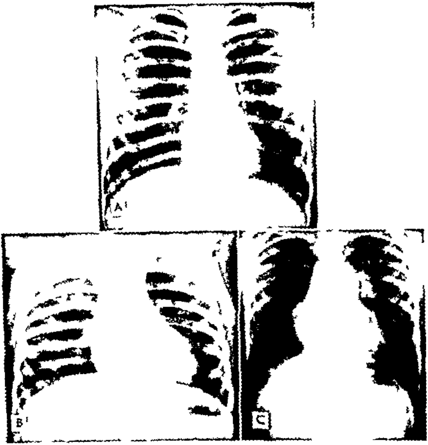


Figure 28 (A) Normal pulmonary vasculature (B) Pulmonary vascular engorgement (C) Pulmonary ischemia

One further peculiarity of the pulmonary vasculature has to be considered here namely the collaterals The question boils down to this Where *do* the pulmonary vessels come from? If they originate as they should from the left and right main pulmonary arteries then these principal vessels should be readily visible in the hilus and the smaller *vessels should diminish gradually from the hilus toward the outer third* Conversely if the pulmonary vasculature is principally made up of bronchial collaterals such as are seen in pulmonary or tricuspid atresia the vessels will not originate from a well defined hilar "comma and their distribution will extend in a lacelike pattern evenly throughout the lung fields Figure 29 shows a predominantly bronchial type of pulmonary vasculature Unequal distribution of the pulmonary vasculature may be seen in certain congenital anomalies involving atresia *of* the left or the right main pulmonary artery

CHAPTER 3

Electrocardiography

Electrocardiography, accurate physical examination and radiology form the tripod on which rests the clinical diagnosis in pediatric cardiology. Omission of, unfamiliarity with or misinterpretation of any of these three tools spells disaster.

This chapter on electrocardiography can perhaps best be introduced by enumerating the things it does not aim to do. (1) It is not a complete treatise on electrocardiography. Excellent textbooks are available for this purpose.[60, 205, 301] (2) It does not make the assumption that the reader knows nothing about electrocardiograms. It is unlikely that an appreciable number of medical school graduates lack this knowledge. (3) It does not describe conditions of no significance in pediatrics. This would be contrary to the purpose of the book.

In a more positive way, the aim is to present a concise and practical discussion of electrocardiograms of children with emphasis on the normal electrocardiogram as well as on the pathologic conditions prevalent in infancy and childhood.

THEORETICAL CONSIDERATIONS

The Electromyogram

Before the electrocardiogram can properly be understood, certain relatively simple physiologic explanations pertaining to the isolated muscle strip are necessary. The inside of the cell, schematically presented in Figure 31, carries a negative charge, the outer surface a positive one. This potential difference has been measured by micro electrodes and has been found to be in the neighborhood of 90 millivolts. If a galvanometer is connected to two points on the surface of a resting muscle no potential difference is registered, since all points on the exterior of the unstimulated cell have the same potential (Fig. 32 *A*). If the strip is stimulated, the potential on the surface of the membrane drops to zero at the stimulated point, because the membrane becomes electrically permeable and the charges on the two sides neutralize each other. As the wave of excitation spreads, the entire surface of the muscle reverses its positive charge. If a galvanometer is connected to two points on the surface of a stimulated muscle, then, in contrast to the resting state, potential differences are registered: the already depolarized portion of

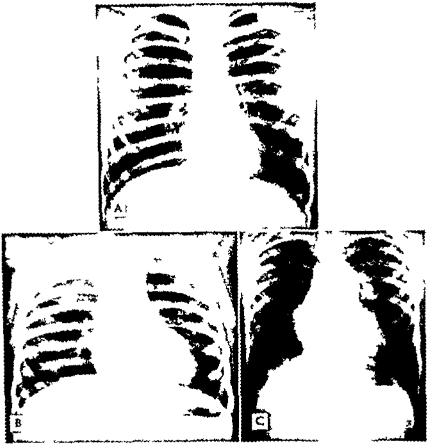


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Figure 29 Extensive collateral circulation in patient with pulmonary atresia

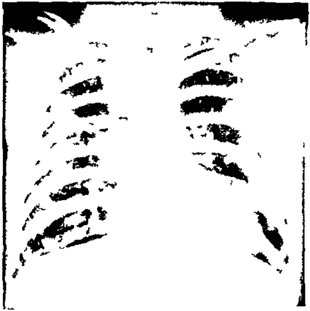


Figure 30 Rib notching in patient with coarctation of the aorta

Dilated intercostal arteries may exert sufficient pressure on the lower end of the ribs to erode them giving rise to "rib notching" pathognomonic of coarctation of the aorta (Fig 30)

ORTHODIAGRAPHY

Orthodiagraphy consists in the tracing of the cardiac silhouette on the fluoroscope screen without distortion by means of a "single" roentgen beam The tracing is reproduced on a piece of onion skin paper attached by clips to the screen This method is accurate and inexpensive and leaves a permanent record thus combining the advantages of fluoroscopy and radiography The principal disadvantages are that it is time-consuming and occasionally it exposes the patient to more radiation than is desirable Within recent years I have had no experience with this method If no equipment for radiography is available fluoroscopy combined with orthodiagraphy should be used Excellent descriptions of the technique are available elsewhere [17]

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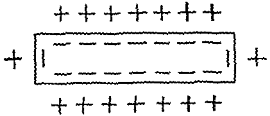


Figure 31 Resting muscle fiber (schematic drawing) showing the negative charges inside and the positive charges outside the cell

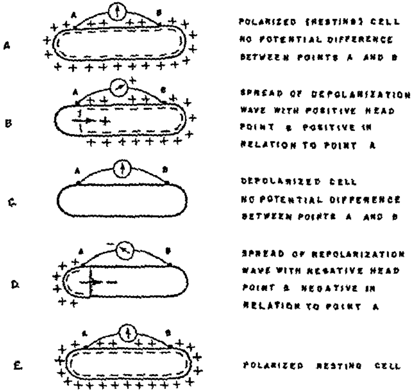


Figure 32 Electrical phenomena in single muscle fiber stimulated at one end

the muscle is electrically negative compared to the intact as yet untouched portion (Fig 32 *B*) After the wave of excitation with a positive head has completely covered the cell membrane there remains no potential difference between the inner and outer surfaces of the entire muscle it is as if the entire electrical energy of the strip had been fired off or depolarized (Fig 32 *C*) As soon as the depolarization is completed the muscle strip starts to "refuel" to repolarize during this process the membrane regains its insulating capacity—the outer surface obtains a positive charge whereas the inside becomes negative The

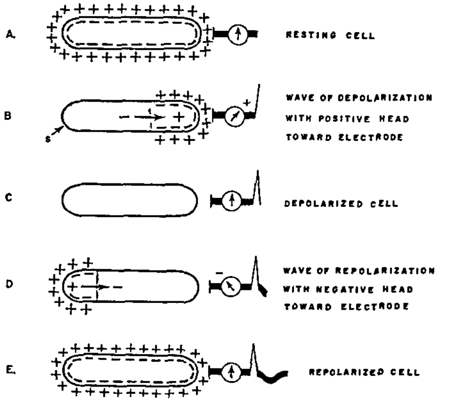


Figure 33 An electromyogram registered from a galvanometer set up at the end of a single muscle fiber at the pole opposite the point of stimulation

spread of this "repolarization wave" is in the same direction as the de polarization wave only as shown in Figure 32 *D* the head is now negative instead of positive With the completion of repolarization the muscle returns to the resting state (Fig 32 *E*)

If a galvanometer is connected to the end of the muscle strip in such a manner that the positive potentials are registered as an upward deflection and the negative ones as a downward deflection then the depolarization and repolarization waves are described as a diphasic curve the so-called electromyogram (Fig 33)

One final word about the physiologic background According to recent observations the changes in membrane potential during depolarization may be defined chemically to a large extent by the exodus of potassium ions from the inside of the cell Conversely repolarization according to this theory may be explained by a re entry of potassium from the extracellular fluid into the interior of the cell after which it is sealed in by the membrane

The Electrocardiogram

By connecting a galvanometer to the heart an electrocardiogram may be obtained This in contrast to the simple diphasic curve of the

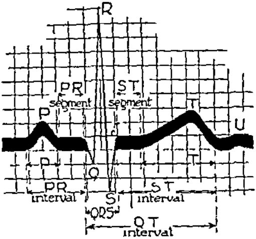


Figure 34 The waves of the normal electrocardiogram (After Burch and Winsor A Primer of Electrocardiography Philadelphia Lea & Febiger)

myogram consists of three elements—P QRS and T—all of which com monly point in the same direction (Fig 34)

It cannot be emphasized enough that the electrocardiogram represents the action current of electrical happenings in the heart and bears very distant relationship to mechanical forces As a matter of fact elec trical phenomena precede in time the muscular contraction hence the electrocardiogram cannot reflect the effective force of the contraction

The principal differences between the myogram of a striated muscle and the electrocardiogram probably are due to four main factors (1) the difference in muscle refractory period (2) the syncytial nature of the heart muscle (3) the presence of a rather complicated conduction sys tem and (4) the problems in placing the galvanometer in the vicinity of the heart

The refractory period of the heart muscle is considerably longer than that of striated muscle This fact is responsible for the inability of the cardiac muscle to go into tetanic contraction it enables the heart to beat rhythmically and to rest between contractions The striated muscle can go into an "oxygen debt" during prolonged continuous stimulation a debt that can be paid during a prolonged resting period The heart for obvious reasons cannot get a prolonged rest hence the long refractory period furnishes it with time to "refuel between beats

The syncytial nature of the heart muscle is responsible for the fact that the electrocardiogram in contrast to the myogram is not a diphasic curve The wave of depolarization of the ventricles as represented by the QRS complex is not followed by a mirror image repolarization wave but rather by a lower and wider most commonly upward wave—the T wave The deviation of the electrocardiogram from the simple diphasic pattern of the muscle strip is called the ventricular gradient and pre

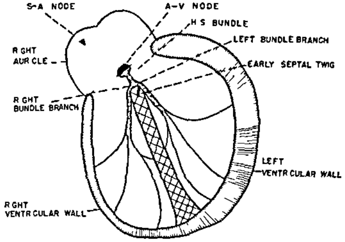


Figure 35 The cond ct on system of the l eart (After Wolff[381])

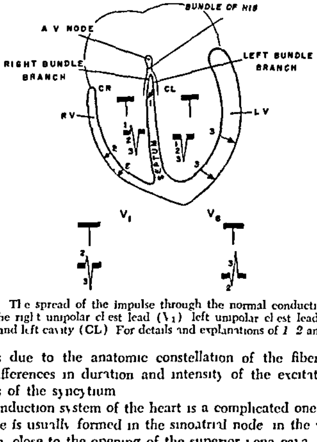


Figure 36 Tl e spread of the impulse through the normal conduction system as reflected at the rigl t unipolar cl est lead (V_1) left unipolar cl est lead (V_6) rigl t cavity (CR) and left cavity (CL) For details and explanations of *1* *2* and *3* see text

sumably is due to the anatomic constellation of the fibers and the resulting differences in duration and intensity of the excitation in different parts of the syncytium

The conduction system of the heart is a complicated one (Fig 35) The impulse is usually formed in the sinoatrial node in the wall of the right atrium close to the opening of the superior vena cava From here it spreads like a ripple in a pond through both atria the right atrium is activated first the left atrium later As the wave of excitation spreads through the atria it reaches the atrioventricular node in the wall of the

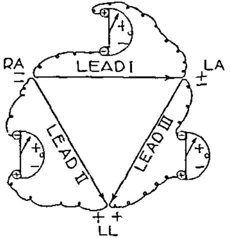


Figure 37 The bipolar leads of Einthoven and their reflection on the galvanometer (After Burch and Winsor[60])

right atrium near the opening of the coronary sinus From the atrio ventricular node the impulse spreads down toward the ventricles through the atrioventricular bundle (His) and its branches Ordinarily the septum is activated from left to right (Fig 36) Next the impulse reaches the narrow wall of the right ventricle for a short time only to be overpowered by the impulse reaching the thicker left ventricular wall the final portion of the depolarization process It should be remembered that the process of depolarization spreads through the ventricular wall from the Purkinje fibers through the endocardium outward toward the sub epicardial layers conversely repolarization probably begins subepicardially and spreads toward the endocardium

The position of the electrode is the fourth variant determining the shape of the electrocardiogram The bipolar leads described by Einthoven (Fig 37) originate from the extremities and register potential differences between extremities—the right arm the left arm and the left foot Lead I represents potential differences between the right arm and the left arm lead II between the right arm and the left foot and lead III between the left arm and the left foot These are the bipolar extremity leads or standard limb leads of Einthoven In contrast the unipolar leads the so called V leads measure what may be considered actual electrical potentials at some given point in the body The unipolar limb leads aV_R aV_L and aV_F[17] (or V_R V_L and V_F)[357] measure the electrical potentials at one extremity Lead aV_R measures the potentials at the right arm aV_L at the left arm and aV_F at the left foot The unipolar chest leads measure the electrical potentials of the heart at certain definite points on the chest By convention the American Heart Association advocates the use of six chest leads V_{1-6} to which in pediatrics three

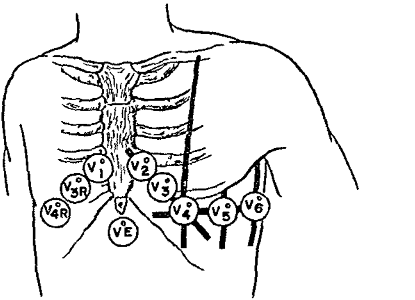


Figure 38 Location of the unipolar chest leads commonly obtained in children

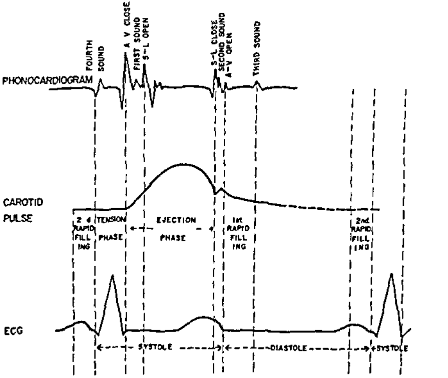


Figure 39 The electrocardiogram and its correlation with the various mechanical activities of the cardiac cycle

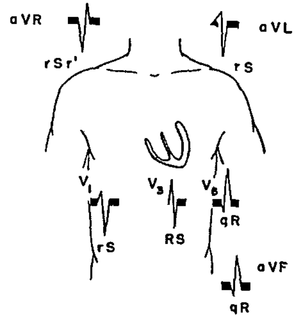


Figure 40 The normal unipolar limb and chest leads in an adult with a vertical heart For details see text.

more (V_{3F} V_{4R} and V_E) often must be added to obtain a clear cut pattern of progression (Fig 38)

Various types of curves are obtained depending on the structure of the heart muscle the attributes of the conduction system and the position and number of the electrodes The basic tracing and its correlation with the activities of various portions of the heart are presented in Figure 39

Certain conventions are observed in the setting of the galvanometer In the standard limb leads where potential differences are explored the galvanometer shows a positive deflection in lead I if the right arm potential is less than that of the left arm in lead II if the right arm potential is less than that of the left foot and in lead III if the left arm potential is less than that of the left foot From these conventions follows the fact that at any one instant in the P QRS T complex the mathematical sum of lead I plus lead III equals lead II As an example if the height of R_1 is +8 mm and the height of R_3 is −4 mm then the height of R must be +4 mm

The conventions ruling the registration of the unipolar leads require that the galvanometer be set in such a way that when the impulse travels *toward* an electrode a positive deflection is described whereas if the impulse travels *away* from it the deflection described will be a negative one Taking the diagram describing the spread of the impulse from the atrioventricular node through the ventricles it is possible to reconstruct

Table 4 Patterns of QRS and T Waves in Various Regions of the Heart [95]

	QRS	T
Surface of right ventricle pulmonary conus	rSr¹	T−
Surface of right ventricle basal (lateral wall)	rS or RS	T±
Surface of right ventricle apical	Rs	T+
Cavity of right ventricle	rSr¹ or rS	T−
Cavity of left ventricle	QS	T−
Atrioventricular border of left ventricle (transition between cavity and surface) back of the heart	Qr or QR	T−
Surface of left ventricle basal (lateral wall)	qR or qRS	T+
Surface of left ventricle apical	qR or qRS	T+

the tracings obtained from various unipolar chest and limb leads (Figs 36 40 Table 4) It may be seen that the initial deflection of V_1 representing a unipolar lead from the right chest will be a notched positive one made up partly from the activation of the septum from left to right and partly from a short period of right ventricular activation before the thicker left ventricle asserts itself The final portion of V_1 consists in a deep negative deflection caused by the impulse spreading through the left ventricle away from the right ventricular (V_1) electrode On the other hand if the electrode is placed on the surface of the lateral wall of the left ventricle or to V_6 the electrocardiograph characteristically registers a qR pattern It is demonstrated in Figure 36 that the q is due to the impulse initially traveling away from the left ventricle activating the septum and later the right ventricle from left to right The tall R ensuing represents the impulse spreading through the bulky left ventricle toward the electrode at V_6 A third pattern obtainable from the surface of the medial aspects of the left or the right ventricle or from chest lead V_{3-4} is the so called transitional zone pattern RS This is the result of the galvanometer s registering both left and right ventricular potentials with equal deflections A pattern usually obtained from lead aV_R is the right ventricular cavity pattern rSr¹ representing the electrical forces from the back of the heart Most of the impulses except those from the very terminal portion of the left ventricle are directed away from this electrode hence the predominantly downward deflection Patterns not ordinarily obtained in the unipolar leads of normal adults are *Rs which represents right ventricular surface potentials and is characteristic of right ventricular hypertrophy* and QS which is obtainable from inside the left ventricle through intracardiac electrocardiography

The electrical position may be calculated from the unipolar lead patterns enumerated (Fig 41) If a left ventricular pattern (qR) is registered by the left arm lead the position is a horizontal one If this pattern is obtained in the left foot lead the position is vertical If both aV_F and aV_L show a qR pattern the position is called intermediate

The mean electrical axis may be calculated from the standard bipolar leads by plotting the magnitude and direction of the main QRS complexes in leads I and III on a diagram Vertical lines are then drawn

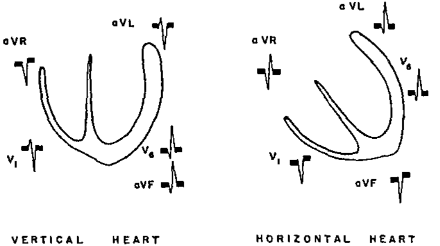


Figure 41 The electrical position of the heart calculated from the unipolar leads For details see text

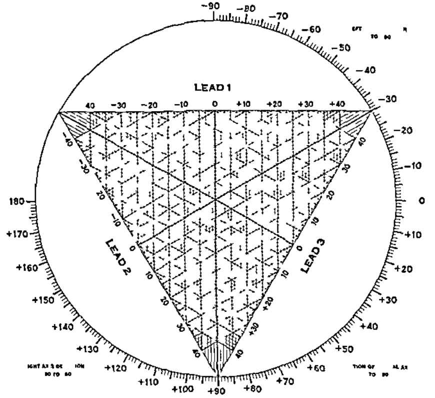


Figure 42 Diagram for calculating electrical axis (From Sanborn Company Cambridge Massachusetts)

from the end point in leads I and III The intersecting point of these two lines connected with the center of the diagram gives the direction in degrees of the mean electrical axis (Fig 42) If the direction of the main complex in lead I is negative and that of lead III positive there is right axis deviation (+90 to +180 degrees) Conversely tall R_1 and deep S_3 means left axis deviation (0 to —90 degrees) Normal axis is between 0 and +90 degrees

It should be emphatically mentioned here that deviation from the normal axis may be the result either of ventricular hypertrophy or of position Axis deviation per se should not be considered equivalent to ventricular hypertrophy It should also be pointed out that infants and children as a rule show right axis deviation as a result of both vertical position and right ventricular dominance

PRACTICAL ELECTROCARDIOGRAPHY

How to Take an Electrocardiogram on a Child or an Infant

The technique of obtaining an electrocardiogram in an older child should present no particular problem The techniques described in any handbook by any reputable manufacturer of cardiographs should contain sufficient instructions

Infants on the other hand present considerable often insurmountable problems In my experience direct writing machines are infinitely more practical than photographic ones in the electrocardiography of infants The fact that the baby is not likely to hold still for any length of time makes it imperative that the technician see instantaneously whether a few satisfactory complexes without somatic interference have been obtained in any one lead With a photographic machine even after a great deal of experience it is often impossible to tell prior to developing the film whether a satisfactory tracing has been obtained I am well aware of the magnificent records obtained on a two channel photographic machine by Doctor Ziegler[369] of Detroit I can only express admiration for his ability and at the same time express doubts that such tracings could be obtained in large numbers in the majority of institutions

Only a technician who likes children is likely to obtain clear tracings the reassurance and kindness emanating from such a person may make a tremendous difference A bottle of milk or a lollipop may quiet a restless infant at other times the only way a satisfactory record can be obtained is to have the mother hold the baby in her lap

In infants the smallest obtainable electrode should be used a chest piece with a ½ inch diameter usually suffices

It is important that the electrode paste should touch the chest only at the place where the particular lead is if the paste is smeared across the chest a single pattern may be obtained from V_{1-6}[71]

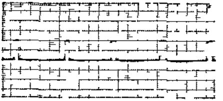


Fig 43

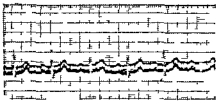


Fig 44

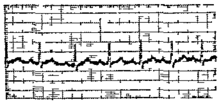


Fig 45

Figure 43 Normal standardization
Figure 44 Sixty cycle interference
Figure 45 Somatic tremor

Finally in an infant with a small chest lead V_3 may be omitted from the series of chest leads On the other hand V_E (ensiform process) and V_{4R} or V_{3P} should be taken routinely

General Evaluation

The general evaluation of the tracing should include adequate evi dence of correct standardization a current of 1 millivolt introduced instantaneously into the machine should produce an excursion of 10 mm in height and the upstroke time (interval between onset and zenith of the upward deflection) should be less than 0 02 second (Fig 43)

The base line should be free from regular 60 cycle interference (Fig 44) and from irregular somatic tremors (Fig 45) A wandering base line or senseless and irregular deflections are usually due to faulty application of the electrodes

Determination of Rate and Rhythm

This should begin by identification of the P and QRS complexes usually in lead II or in whichever standard limb lead they are outlined

Table 5 Rate per Minute by Electrocardiogram Interspaces between 2 QRS Complexes

5	300	13	115	21	72	29	52
6	250	14	107	22	68	30	50
7	214	15	100	23	65	31	48
8	188	16	94	24	63	32	47
9	167	17	88	25	60	33	45
10	150	18	84	26	58	34	44
11	136	19	79	27	56	35	43
12	125	20	75	28	54		

most clearly Next the relationship between the QRS complex and the P wave must be determined in terms of constancy or variability If the relationship is constant the ventricular rate per minute is counted if the relationship is variable both atrial and ventricular rates must be determined The determination of the rate can most easily be accomplished either by counting the number of small squares between two consecutive QRS or P complexes (Table 5) or especially if the rhythm is irregular by multiplying by 20 the number of cycles between two vertical lines (three seconds) on standard electrocardiograms

The P QRS T U Complex

The P Wave The P wave the first complex in any cardiac cycle results from the electrical activity of the atria (Fig 39) There is evidence that the first part of this wave is the result of the activity of the anterior portion of the right atrium whereas the final part corresponds to the depolarization of the left atrium and the posterior part of the right atrium The duration of the P wave in children measured in standard limb leads averages 0 06 second (standard deviation equals ±0 02) [70] *Its maximum height is less than 2 5 mm in any lead* [8, 369] The shape of the P wave is usually rounded in the standard limb leads maximal in lead II and usually minimal or even inverted (2 per cent of normals) in lead III The right chest leads usually show diphasic or inverted P waves with the peak appearing early P in V_5 and V_6 in contrast is usually upright but low with a relatively late peak P in aV_R is normally inverted in aV_L is upright

It may be said with fair certainty that P waves of more than 2 5 mm in height or more than 0 08 second in duration are abnormal in children and denote atrial hypertrophy Some differentiation between right and left atrial hypertrophy is usually though not invariably possible Peaked or "gothic shaped" tall but not prolonged P waves in leads II and III coupled with pointed P waves in chest leads V_1 usually indicate right atrial hypertrophy (P pulmonale) (Fig 46) In contrast flat topped P waves in leads I and II of normal height but prolonged duration with two notches at least 0 05 second apart indicate left atrial hypertrophy (P mitrale) (Fig 47) In the chest leads promi

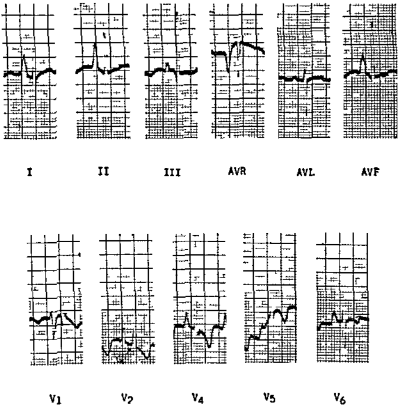
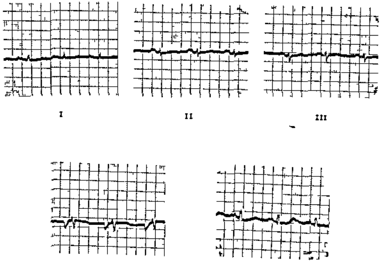


Figure 46 Giant P pulmonale in patient with pure pulmonary stenosis Note tall thin Gothic P wave in lead II and a mirror image of this in aV also tall pointed P in aV and V_4



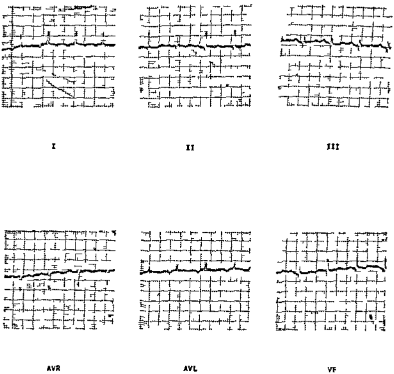


Figure 48 Inverted P waves in patient with dextrocardia Note downward P in lead I upright P in lead II and upright P in aV inverted P in aV_L

nent and late P waves may be seen in V_5 and V_6 British authors[337] maintain that combined atrial hypertrophy may be recognized on the basis of the presence of both tall and broad P waves the tall early ones being visible in the right chest leads the late ones in the left chest leads

Absence of P waves in all leads means absence of atrial activity It is wise however to explore the regions above V_1 and V_{4R} with a unipolar electrode and perhaps even to obtain an esophageal lead before the absence of P waves is diagnosed with certainty Occasionally weak atrial activity is not registered in the conventional leads

Inverted P waves in lead I (Fig 48) indicate dextrocardia and *obviously should be accompanied by upright P waves in* aV_R *and* inverted P waves in aV_L A P wave of unusual shape with a P R interval of less than 0 10 second usually means that the pacemaker is not in the sinoatrial node hence the designation "ectopic atrial" or "nodal" pacemaker

The normal or nodal P waves may be replaced by bizarre forms of atrial activity known as atrial flutter or fibrillation In atrial flutter the atrial rate is about 200 to 300 beats per minute the complexes are regular and triangular in shape in leads II and III but barely visible in lead I (Fig 49) The flutter waves are continuous and may be followed through the QRS complexes Atrial fibrillation is the severest disturbance of atrial

I

II

III

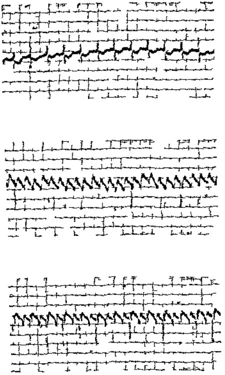


Figure 49 Atrial flutter Note the regular triangular shaped complexes in leads II and III with almost no visible atrial activity in lead I

Table 6 Maximal P-R Intervals in Seconds at Different Age Levels and Varying Heart Rates

Age	*Rate*					
	< 71	71–90	91–110	111–130	131–150	> 150
<1 mo			0.11	0.11	0.11	0.11
1–9 mos			0.14	0.13	0.12	0.11
10–24 mos			0.15	0.14	0.14	0.10
3–5 yrs		0.16	0.16	0.16	0.13	
6–13 yrs	0.18	0.18	0.16	0.16		

From M M Alimurung and B F Massell The Normal P-R Interval in Infants and Children *Circulation 13* 257 1956

activity has a rate of more than 350 beats per minute and is completely irregular (*Fig.* 50)

The P-R Interval *This should be measured from the beginning of* the QRS complex preferably where a clear-cut Q wave is visible Careful study of a large number of electrocardiograms in adults [8] resulted in

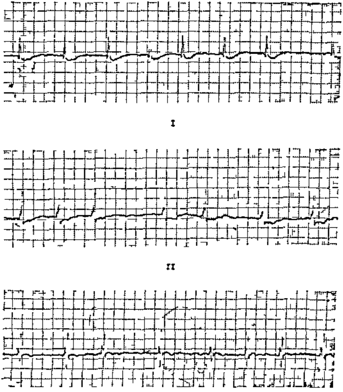


Figure 50 Atrial fibrillation Note the irregularly spaced QRS complexes the absence of well defined atrial activity in lead I and slight coarse fibrillary waves in leads II and III

the concept that the longest P R interval found in any limb lead should be taken as the correct value for atrioventricular conduction time

When determining the P R interval in children both the rate and the age have to be considered (Table 6) whereas in the adult only the rate has to be taken into consideration By and large the P R interval decreases with rate and increases with age However the increase in P R interval in older children is greater than one would expect from the slowing of the pulse rate alone

Unusually short P R intervals (less than 0 10 second) may be seen (1) in infants less than one year of age (2) if the pacemaker is not the sinoatrial node or (3) if the atrioventricular conduction is through an aberrant short pathway (Wolff Parkinson White syndrome)[370] (Fig 51) Prolonged P R intervals may be seen in active carditis but are not diagnostic of that disorder Permanently prolonged P R intervals may be congenital (with or without congenital heart disease) or may be the consequence of past rheumatic infection without implying rheumatic activity at all

Regularly variable P R intervals may be seen in the so-called

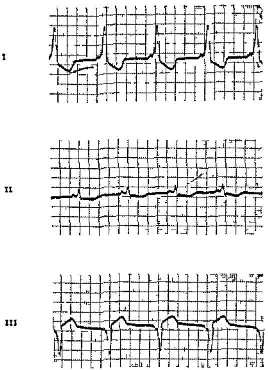
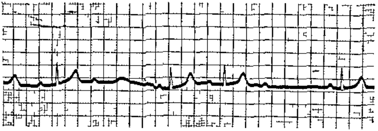


Figure 51 Wolff Parkinson White syndrome Note the short P R interval and the wide QRS complex with T waves in a direction opposite to the main QRS complex



Lead I

Figure 52 Wenckebach phenomenon Note that the second P wave in the tracing is not followed by a QRS complex The third and fourth P waves are followed by QRS complexes but the P R interval gradually lengthens and the fifth P wave is again completely blocked

Wenckebach phenomenon[3] (Fig 52) and in interference with dissociation (Fig 53) Completely irregular P R intervals mean complete atrioventricular block (Fig 54)

The QRS Complex The first downward wave beyond the P wave has been designated as the q wave The first positive deflection beyond the atrial wave is called R and the negative potential following the first

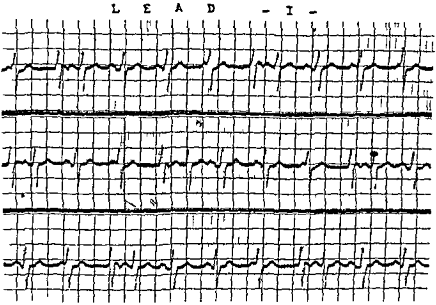


Figure 53 Interference and dissociation Note the regularly spaced P waves and the regularly spaced QRS complex The ventricles are beating somewhat faster than the atria thus the P R interval consistently shortens When the P wave falls behind the QRS complex this stimulates a second QRS complex immediately thereafter In the top row the P R interval shortens progressively from the third P wave through the fourth and the fifth The sixth P wave occurs after the QRS complex and the next QRS complex comes at a short P R interval after the seventh P wave

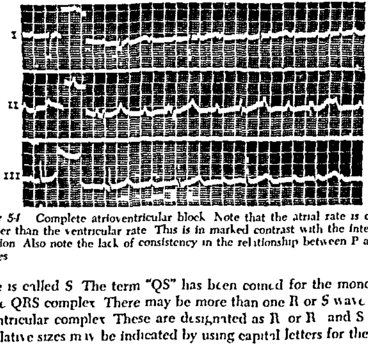


Figure 54 Complete atrioventricular block Note that the atrial rate is considerably faster than the ventricular rate This is in marked contrast with the interference dissociation Also note the lack of consistency in the relationship between P and QRS complexes

R wave is called S The term "QS" has been coined for the monophasic negative QRS complex There may be more than one R or S wave within one ventricular complex These are designated as R or R and S or S their relative sizes may be indicated by using capital letters for the larger

Table 7 Maximal QRS Voltage in the Chest Leads[a]

Age Group	Lead	Q Wave Mean	Q Wave Min	Q Wave Max	Q Wave 10%	Q Wave 90%	R Wave Mean	R Wave Min	R Wave Max	R Wave 10%	R Wave 90%	S Wave Mean	S Wave Min	S Wave Max	S Wave 10%	S Wave 90%
I	V	0	0	0	0	0	10 3	2	2	5	17	4 95	0	23	1	1
	V	0	0	0	0	0	15 6	5	33	9	23	10 5	0 5	28	3	19
	V	0 18	0	2 5	0	0 5	16 7	4	36	10	24	11 3	0 25	25	5 75	17
	V	0 6	0	3 5	0	2 5	13 1	2 5	28	7 5	3	5 74	1	15	2	10
	V	0 83	0	3 5	0	2	9 3	0 25	4	1	16	3 12	0	11	0 5	7
II	V	0	0	0	0	0	8 23	0 5	26	3 5	13	6 03	0 5	22	1 5	13
	V	0	0	0	0	0	15 3	3	30	8	3	14 2	0 25	28	7	23
	V	0 17	0	5	0	0 5	17 9	6	30	10 75	26	11 4	2	25	5	18
	V	0 08	0	5	0	3	14 6	4	8	6	21	4	0	15	1 5	7
	V	1 26	0	5	0	2	10 6	2 5	4	5	15	1 6	0	6	0 25	3 5
III	V	0	0	0	0	0	8 5	0 5	20	2 5	11	9 8	1	25	3	17
	V	0	0	0	0	0	12 5	3 5	25	6	19	17 3	4	33	8	7
	V	0 12	0	3	0	0 5	18 3	5 5	37	10	8	11	1	34	5	17
	V	1 25	0	9	0 5	3 5	19 5	7	40	11	25	3 35	0	13 5	0 75	6 25
	V	1 28	0	8	0 25	2 5	13 7	4	29	8 25	19	1 06	0	6	0	2 5
IV	V	0	0	0	0	0	4 9	0 5	14		9	10 7	1	27	4 5	18
	V	0	0	0	0	0	9 7	2	1 8	5	14 5	19 5	6	35	11	7 5
	V	0 13	0	2	0	0 5	18 4	3	37	11	49	10 7	0 25	24	4	19
	V	1	0	4	0 5	2	19	5	37	13	6	2 76	0	14	0 25	6
	V	1 04	0	4	0 25	2	13 9	4 5	33	8 5	20	0 9	0	8	0	3

Note 10% column indicates voltage [illegible] 10 per cent of cases below the minimum and 90% column indicates voltage [illegible] 90 per cent of cases below the minimum

deflections and small letters for the smaller ones This type of detailed description is usually used only in unipolar leads

The QRS complex is the depolarization wave of the ventricles Normally its duration is less than 0 10 second the average duration increases only slightly with age (0 065 second at twenty four hours to 0 075 second at twelve years of age)[369] and is influenced relatively little by rate A QRS complex of 0 10 to 0 12 second in duration is called incomplete bundle branch block a duration of more than 0 12 second is complete bundle branch block The measurement should be made in lead II or in one of the chest leads and should be started with the onset of the Q wave and terminated where the electrocardiogram line returning from S toward the base line suddenly thickens The longest QRS in a tracing should be measured

The q wave has no particular significance in pediatrics Its vital role in the diagnosis of myocardial infarction in adults is well known It is not commonly found in lead I but is frequently present in leads II III aV_F and V_5 and V_6 A q followed by R is never found normally in V_{4R} or V_1 a QS in the right chest leads on the other hand is not especially uncommon in healthy children Occasionally in cases of right ventricular hypertrophy a qR is present in V_1 close inspection and special techniques often show this deflection to be not a q but in fact an rs wave The duration and the size of the q wave are of no particular importance in children in adults the size should always be less than 25 per cent of the amplitude of the succeeding R wave

The R wave is the major positive deflection in the electrocardio

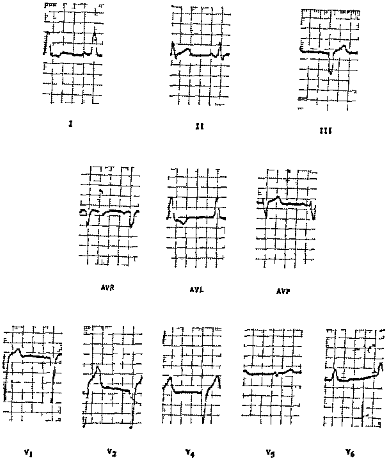


Figure 55 Left bundle branch block Note tall and wide R_1 deep and wide S_3 and absence of Q in V_6

gram It is usually tallest in the standard limb leads in aV_F and in V or V_6 Certain arbitrary limits of the normal R wave based on the statistical analyses of large numbers of tracings have been drawn up[8, 53, 314, 315, 369] These figures have been useful in diagnosing ventricular hypertrophy but it is to be remembered that all such limits are purely arbitrary and they cannot with certainty determine whether or not any individual deflection is of normal amplitude They can indicate only whether it is likely to be normal or not With these reservations in mind the maximal QRS waves in our study of the chest leads of 521 normal infants and children are reproduced in Table 7

The calculation of the mean electrical axis on the basis of the maximal R waves in the standard limb leads and the determination of the electrical position of the heart based on the pattern of the unipolar limb leads have been discussed before (p 50)

One other measurement of the R wave deserves mention This is the so called intrinsicoid deflection designated as "ventricular activation

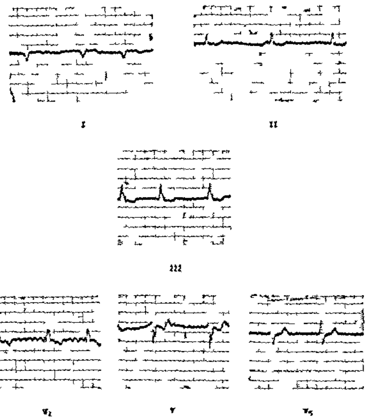


Figure 56 Right bundle branch block Note deep and wide S in lead I and tall and wide R lead III splintered upright V_1 and deep S V_5 This patient also has atrial fibrillation as noted in lead V_1

time" (VAT) This is calculated as the time interval between the beginning of the q wave and the peak of the latest R wave (R or R) in any one lead On the basis of certain theoretic considerations the ventricular activation time is thought to be a measure of the thickness of the portion of the ventricular wall underlying the particular unipolar lead where it is being calculated The normal ventricular activation time for the right ventricle is less than 0 03 second for the left ventricle less than 0 04 second

The shape of the R wave gives information about the intraventricular conduction notching or slurring of the R waves means intraventricular conduction disturbance Fine notching may not necessarily be abnormal coarse notchings are almost invariably pathologic Bizarre and *wide (more than 0 12 second)* QRS *complexes are seen with complete bundle branch block* In the standard limb leads of left bundle branch block (Fig. 55) left axis deviation is present and R_1 is tall and late and commonly followed by a sharply inverted T wave In contrast

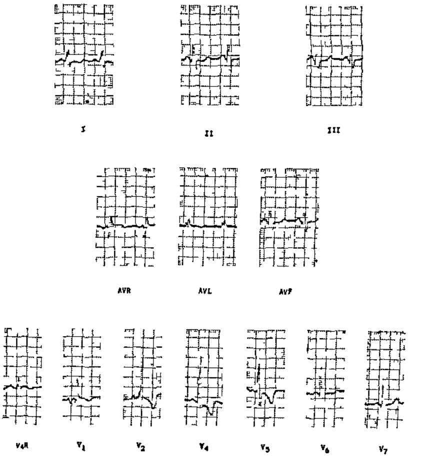


Figure 57 Incomplete right bundle branch block Note the splintered R in lead I V_1 and lead II This patient also had right ventricular hypertrophy as exemplified by the tall R in V

in right bundle branch block right axis deviation is seen S_1 is wide and deep and T_1 is upright (Fig 56) The QRS complexes of bundle branch block are identical with those of ventricular premature beats or ventricular tachycardia but the relation of the P wave to the QRS complex is different in the two conditions There is a fixed and regular P QRS relationship in bundle branch block whereas ventricular premature beats or ventricular tachycardia is completely independent of atrial activity

The appearance of secondary R waves (R or R) in the chest leads with or without QRS prolongation is indicative of incomplete bundle branch block As has been mentioned a QRS complex of supraventricular origin with a duration of 0 10 to 0 12 second indicates incomplete bundle branch block irrespective of its configuration If the secondary R is noted in the right chest leads incomplete right bundle branch block is present (Fig 57) if it is found in V_{5-6} it means incomplete left

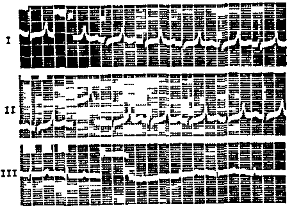


Figure 58 Potassium intoxication with tall tent shaped T leads I and II

bundle branch block[301] Incomplete right bundle branch block especially with small secondary R waves is present in a high percentage of perfectly normal infants and in some children

The S wave is usually deepest in leads I aV_R and V_1 Arbitrary limits of the type mentioned in respect to R waves have also been drawn up for S waves in the chest leads (Table 6) Normal limits of S_{V1} plus R_{V5-6} have also been calculated[8, 315] Wide S_1 and S_{V5-6} are characteristic of complete or incomplete right bundle branch block Conversely wide S_3 and $S_{V1,2}$ are seen with left bundle branch block Deep S_1 indicates right axis deviation

The RS T Segment The RS T segment is the interval between the end of the QRS complex and the beginning of the T wave In most instances it is difficult to separate it clearly from the T wave proper since they are both part of the same process of repolarization The duration of the RS T segment is almost impossible to determine Under ordinary circumstances it is at the level of the T P segment depressions or elevations of 1 5 mm or more are probably pathologic and indicate myocardial ischemia or digitalis effect Marked prolongation of the S T segment suggests electrolyte imbalance specifically hypocalcemia

The T Wave The T wave the terminal portion of ventricular activity is always positive in lead I Negative T suggests myocardial damage negative T_3 is within normal limits Inverted T_1 suggests either dextrocardia or severe myocardial impairment (ischemia electrolyte disturbance and so forth) T in aV_R is always inverted T in aV_L or aV_F is variable T_{V5-6} is always upright in normal persons except during the first few hours of life In contrast inverted $T_{V1,3}$ is normal in infants and children beyond the first week of life

Tent shaped peaked T waves are characteristic of potassium intoxication (Fig 58)

The Q T Interval The Q T interval is the measure of total electrical systole of the ventricles (Fig 39) Its duration is dependent much

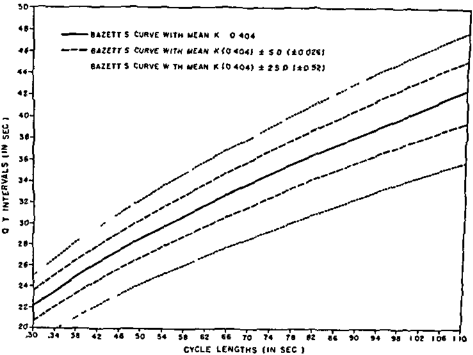


Figure 59 Calculation of the corrected Q T interval from the actual Q T measurement and the cycle lengths in seconds (From Alimurung Joseph Craig and Massell [7])

like that of the P R interval on age and rate Much controversy surrounds the measurement and the significance of the Q T interval [7, 330] Conventionally it is measured from the beginning of the QRS complex to the end of the T wave Lepeschkin[204] conclusively proved that in a certain number of cases the measurement thus performed involves the U wave as well *In any determination of the Q T interval the averages of at least three complexes obtained from three different leads, should be measured* The figure thus obtained should then be compared with the table of expected normals for age and rate [15] Alimurung and his associates [7] using Bazett s formula $\left(U = \frac{Q\,T}{\sqrt{R\,R}}\right)$ [9] constructed a useful graph for the determination of the normal Q T interval in any given cycle length in children this graph includes a mean curve and curves to twice the standard deviation the latter including 95 6 per cent of the normals (Fig 59) Prolonged Q T interval is present in a certain number of cases of carditis[130] and in some instances of electrolyte imbalance (potassium or calcium depletion) Shortened Q T interval may be the result of digitalis administration According to Taran it also occurs as the consequence of pericarditis [330]

The *U* Wave *The U wave is mentioned only for the sake of completeness* Its practical importance is not known The U wave is a

broad low wave discernible in the standard limb leads of about half of the normal electrocardiograms It may be prominent if the cardiac output is high or if the patient is under the influence of drugs (insulin digitalis) or it may accompany changes in cell chemistry (potassium calcium)

NORMAL UNIPOLAR PRECORDIAL AND EXTREMITY LEADS IN INFANTS AND CHILDREN

The normal adult left ventricle is thicker than the right ventricle This anatomic difference is probably the consequence of the pressure differences between the systemic and pulmonary circuits As a result of these anatomic and physiologic facts the normal adult electrocardiogram is characterized by left ventricular dominance This may best be seen in the unipolar chest leads where as mentioned before the typical pattern over the right ventricle is rS whereas the left ventricular leads show a qR or Rs wave (Table 4 Fig 40) This is referred to as normal adult R/S progression from V_{1-6}

In contrast to that of the adult the circulation of the newborn infant is characterized by anatomic and physiologic dominance of the right ventricle These facts are clearly mirrored in the unipolar chest leads where distinct differences between infants and adults are noted As expected the unipolar chest leads of the newborn infant show right ventricular dominance or hypertrophy namely right chest leads V_{4R} to V show a dominant R wave (Rs) in all instances[8] The left chest leads show a dominant S (rS) a complete reversal of the adult R/S progression in 50 per cent of the cases The differences between the unipolar chest leads of adults and infants are shown in Figure 60 The probable spread of the impulse in the heart of a newborn child is demonstrated in Figure 61

Between these two extremes the heart of the newborn infant and the heart of the adult the rest of infancy and childhood represents a transitional period from right ventricular to left ventricular dominance All that one may state with certainty is that the unipolar chest leads of normal newborn infants should represent right ventricular dominance and those of adults left ventricular dominance Whether any particular pattern between these two periods of life is normal or not can be expressed only in terms of probability The progression from the newborn to the adult pattern is seen in Figure 62 in which the algebraic sum of R and S waves in V_1 is pictured It may be seen that the sum is a positive one at birth (representing a dominance of R) and gradually becomes a significantly negative one (dominance of S) at thirteen years of age

Lepeschkin[95] stated the problem in another way by saying "The ratio of R to S over the right ventricle the degree of right axis deviation and the thickness of right ventricular wall in percentage of left show an almost identical course during growth"

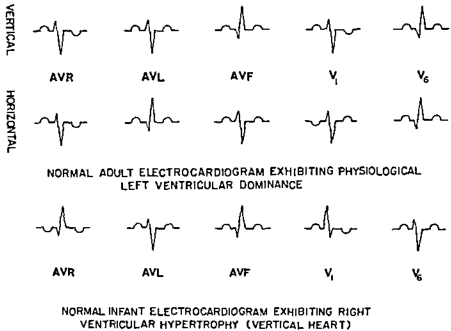


Figure 60 Schematic representation of differences in the unipolar leads of adults with vertical and horizontal hearts and child with vertical heart. Note the dominant R in V_1 in the lowest tracing with deep S V_6 in the same tracing. Also note the tall R in aV_R in this infant tracing. Compare with the deep S in aV_R in both the adult tracings as well as the deep S in V_1 and the tall R in V_6

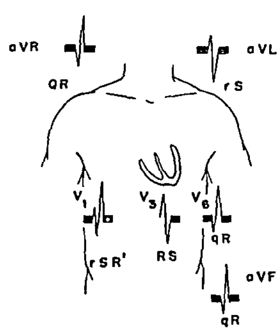


Figure 61 Demonstration of the normal unipolar leads in a newborn infant

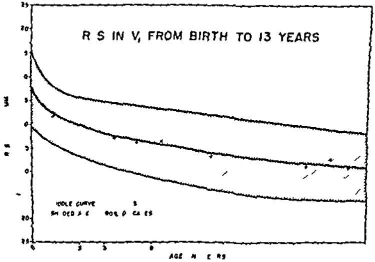


Figure 62 The algebraic sum of R and S waves in lead V_1 from birth to 13 years (From Alimurung Joseph Nadas and Massell [7])

A more detailed approach to the same problem is presented in Table 8 in which 521 infants and children are divided into four age groups and are analyzed according to the pattern of V_1 in their unipolar chest leads It may be seen that whereas 89 per cent of group I show a dominant R 97 5 per cent of group 4 show a dominant S

The T waves in the right chest leads are upright during the first day of life in at least 75 per cent of normal infants T_{V6} on the other hand is inverted in about 50 per cent of infants within the first twenty four hours of life After the first few days inverted T waves in the right

Table 8 QRS Patterns in V_1 in 521 Infants and Children

Group I 0–9 Mos 100 Cases				Group III 3 5 Yrs 124 Cases			
R	2	72 (72%)	89 (89 %)	R	0		
Rs	70			Rs	13		
R > S	17			R > S	10		
R = S	0			R = S	12		
R < S	7			R < S	46	89 (72%)	101 (81 4%)
rS	4			rS	43		
Group II 10 Mos–2 Yrs 98 Cases				**Group IV 6–13 Yrs 109 Cases**			
R	0	76 (77%)		R	0		
Rs	30			Rs	3		
R > S	46			R > S	2		
R = S	0			R = S	15		
R < S	16			R < S	42	179 (89 95%)	194(97 5%)
rS	6			rS	137		

chest leads and upright T waves in the left chest leads are the rule with practically no exceptions In infants T wave inversion may extend through V_{-3} whereas in older children it is restricted to V_1 and only occasionally extends to V_2

The P waves show almost no change throughout infancy and childhood They are almost always inverted or diphasic in V_{1-} and as a rule are upright in V_{5-6}

The unipolar limb leads as discussed previously give valuable information about the position of the heart Seventy five per cent of the infants less than nine months of age have vertical or semivertical hearts This relationship changes but slightly throughout childhood 66 per cent of patients more than six years of age still have vertical or semivertical hearts The horizontal position of the heart is rare in infants less than nine months old

One other significant feature of the young infant s heart is the tall R wave encountered in lead aV_R A dominant R is seen in 28 per cent of infants less than nine months old (almost exclusively in those less than two months old) and in only 2 5 per cent of patients more than six years old

Ventricular Hypertrophy

Perhaps the most important use of electrocardiography in pediatrics is for the determination of the relative thickness of the two ventricles and identification of ventricular hypertrophy

From what has been said in the section on the unipolar chest leads of infants and children it should be clear that a relatively thick right ventricle and thus right ventricular dominance is the rule in infants Conversely a relatively thick left ventricle with left ventricular dominance is invariably found in normal adults (Fig 60) It clearly follows that a dominant left ventricle in an infant indicates abnormal left ventricular hypertrophy and a dominant right ventricle in an adult is proof of pathologic right ventricular hypertrophy Over and above these qualitative differences between infants and adults quantitative estimates of the degree of ventricular dominance may lead to the diagnosis of *left ventricular hypertrophy in some adults and of right ventricular* hypertrophy in some infants

The diagnosis of ventricular hypertrophy often rests on the height of the QRS complexes alone If one realizes however that the voltages registered by any of the conventional leads depend not only on the thickness of the heart muscle but also on the thickness of the chest wall and the distance of the heart from the electrode the inevitable fallacies become clear Also one has to bear in mind the fact that "normal voltages" are based simply on minimums and maximums determined on a large but not infinite number of patients Consequently a warning should be sounded here to the effect that ventricular hypertrophy de



Figure 63 Schematic representation of standard limb leads in patients with left ventricular hypertrophy *A* Left axis deviation *B* Tall R in leads II and III

termined on the basis of voltages alone is not an absolute certainty It simply means that it is unlikely—statistically improbable—that a normal heart would give rise to voltages beyond a certain magnitude but it is not impossible in any one case For this reason electrocardiographic voltages should be regarded only against the background of other electrocardiographic and clinical criteria of heart disease and the diagnosis of ventricular hypertrophy should seldom if ever be based on voltages alone

Left Ventricular Hypertrophy STANDARD LIMB LEADS (Fig 63) (*a*) Left axis deviation (tall R wave in lead I and deep S wave in lead III) of more than −30 degrees indicates left ventricular hypertrophy in 50 per cent of the cases Horizontal hearts however without evidence of left ventricular hypertrophy may show significant left axis deviation as well (*b*) The heights of voltages in standard limb leads are also useful in the diagnosis of left ventricular hypertrophy If the sum of the R waves in leads II and III is more than 45 mm or if the sum of the R wave in lead I and the S wave in lead III is more than 30 mm the presence of left ventricular hypertrophy should be suspected

UNIPOLAR LIMB LEADS (Fig 64) These are not pre eminently useful in diagnosing left ventricular hypertrophy Only the voltages may be utilized in these leads since the shape of the complexes is strongly influenced by changes in position Left ventricular potentials reflected in aV_L or aV_F with R waves of 20 mm or more may indicate left ventricular hypertrophy The main deflection in aV_R is always negative in left ventricular hypertrophy

UNIPOLAR CHEST LEADS (Fig 65) Unipolar chest leads in which the effects of position are somewhat minimized are better indices of ventricular hypertrophy than the limb leads (*a*) The height of R in V_{5-6} and the depth of S in V_1 may be used as criteria of left ventricular hypertrophy if they clearly fall outside the transitional zone if V_1 or V_{5-6} still show RS complexes then positions further to the right or the left must be used as the true potentials of the respective ventricles If the sum of the R wave in V_5 or V_6 and the S wave in V_1 or V is more than 45 mm one may be assured that left ventricular hypertrophy

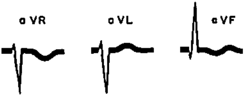


Figure 64 Left ventricular hypertrophy as pictured in the unipolar limb leads showing deep S in aV_R and tall R in aV_F preceded by a q

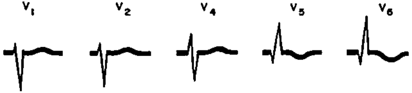


Figure 65 Characteristic unipolar chest leads in left ventricular hypertrophy Note deep S V_1 and V and tall R V_5 and V_6 preceded by q and followed by inverted T

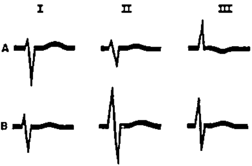


Figure 66 Schematic representation of standard limb leads in right ventricular hypertrophy *A* Right axis deviation *B* Deep S in lead I II and III

exists An R wave in V or V_6 of more than 35 mm is in itself indicative of left ventricular hypertrophy (*b*) An intrinsicoid deflection in V_{5-6} of more than 0 04 second is clear evidence of left ventricular hypertrophy (*c*) Depressed S T and inverted T waves in V_{-6} with or without increased voltages are conclusive evidence of left ventricular hypertrophy

INDIRECT EVIDENCE *Indirect evidence of left ventricular hypertrophy* is furnished by left atrial hypertrophy (Fig 47) (so-called P mitrale) This is in marked contrast to the findings in adults in whom "P mitrale" most often denotes right ventricular hypertrophy secondary to mitral stenosis In children severe organic mitral stenosis is rare thus left atrial hypertrophy usually is the result of a failing hypertrophied left ventricle

In addition to these criteria as emphasized before the finding of a normal adult type of electrocardiogram in a young infant indicates left ventricular hypertrophy

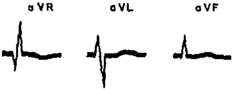


Figure 67 Schematic representation of right ventricular hypertrophy as evidenced in the unipolar limb leads Note the tall late R in aV_R

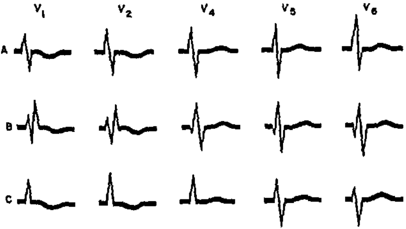


Figure 68 Schematic representation of the different types of right ventricular hypertrophy as pictured in the unipolar chest leads For details see text

Right Ventricular Hypertrophy STANDARD LIMB LEADS (Fig 66) (*a*) Right axis deviation (deep S wave in lead I and tall R wave in lead III) of + 120 degrees or more is suggestive of right ventricular hypertrophy with the usual allowances to be given to positional changes (*b*) A deep S wave in leads I II and III results in an indeterminate axis but strongly suggests right ventricular hypertrophy

UNIPOLAR LIMB LEADS (Fig 67) A dominant R or R in aV_R is characteristic of right ventricular hypertrophy

UNIPOLAR CHEST LEADS (Fig 68) (*a*) The least certain indication of right ventricular hypertrophy in the chest leads consists in marked rotation of the heart resulting in rS or RS deflections from V_1 to V_6 (*b*) An incomplete right bundle branch block pattern in right chest leads consisting of rsR probably indicates right ventricular hypertrophy if the R is more than 10 mm in height The narrower the QRS complex and the taller the R the more right ventricular hypertrophy is suggested A late (more than 0 06 second) R of less than 10 mm in height probably indicates bundle branch block with not much if any right ventricular hypertrophy (*c*) A tall unslurred R_{V1} with or without a q wave and with small if any S wave unquestionably indicates right ventricular hypertrophy (*d*) An intrinsicoid deflection of 0 03 or more

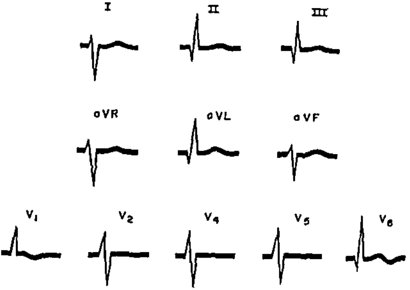


Figure 69 Typical complete 11 lead electrocardiogram in patient with combined ventricular hypertrophy

in V_1 - if not associated with bundle branch block is good evidence of right ventricular hypertrophy (*e*) Inverted T waves in right sided chest leads accompanied by depression of the S T segment in conjunction with the other changes in the chest leads indicate severe hypertrophy ("strain pattern") There seems to be adequate correlation between the severity of the hypertrophy and the extension of the T and S T changes from V_1 toward V_6 (*f*) Positive T_{V1} in infants beyond the first twenty four hours of life suggests right ventricular hypertrophy

INDIRECT EVIDENCE Indirect evidence of right ventricular hypertrophy is furnished by peaked and tall P indicating right atrial hypertrophy ("P pulmonale") (Fig 46)

As emphasized before most of these changes may occur normally in infants but almost never in normal adults The use of the tables in the section on normal chest leads should serve as a guide in determining the significance of these findings in older infants or children (Table 7 *p 61*)

Certain findings even in the youngest infants indicate right ventricular hypertrophy These are (1) the presence of a q wave in V_4 or V_1 (2) an R wave in V_{4R} or V_1 of 20 mm or more without an S wave and (3) S T and T wave changes extending from V_1

The other evidences of right ventricular hypertrophy must be viewed against the background of age and other electrocardiographic and clinical features

Combined Ventricular Hypertrophy (Fig 69) This is usually recognized by one of the following modifications of the left ventricular hypertrophy chest lead pattern (1) a dominant R wave in V_1 (2)

a shift of the transitional zone to the left extending often to V_{5-6} with out obviating the tall R of left ventricular hypertrophy but transferring it to V_7 or V_8 (3) presence of a deep S in lead I or right axis deviation and (4) vertical position

A new concept of ventricular hypertrophy is promoted by the Mexican School of Cardiology particularly by Cabrera In a series of papers Cabrera and his associates[69] proposed an electrocardiographic differentiation between increased flow work (diastolic overloading) and increased pressure work (systolic overloading) of the ventricles Systolic overloading of the right ventricle according to these authors is reflected in the increasingly tall and late R waves with late changes in the S T and T waves in the right precordial leads Diastolic overloading of the right ventricle on the other hand manifests itself in right bundle branch block Our experience supports these views in regard to severe systolic and diastolic overloading of the right ventricle however we also find that mild degrees of systolic overloading may give rise to incomplete right bundle branch block In regard to overloading of the left ventricle I cannot completely support the views expressed by Cabrera and his associates I am unable to diagnose diastolic overloading on the basis of upright T waves in left chest leads as suggested by these authors and I have observed increasing voltages of R_{V5-6} and aV_F with or without changes in the S T and T waves in both systolic and diastolic overload ing of the left ventricle

Arrhythmias

Under ordinary circumstances the rate and rhythm of the heart beat are determined by the activity of the sinoatrial node The average heart rate in resting adults is about 60 to 80 beats per minute In infants it is considerably higher approximately 120 beats per minute in children it is somewhere between these two extremes in the neighborhood of 100 per minute

To determine the "normal rate of any person especially a child the sleeping pulse rate should be counted If a child is awake a number of extraneous factors influence the heart rate to such an extent that it is virtually impossible to obtain a true "resting value

Rhythms Originating in the Sinus Node These are characterized by a P wave of normal and constant configuration and by a P R interval of at least 0 10 second Exceptions to the latter point occur in infants less than one year of age and in patients with the Wolff Parkinson White syndrome[360]

Sinus tachycardia is a rapid regular heart rate ranging to a maxi mum of 180 to 200 beats per minute where the normal relationship between atrial and ventricular complexes is preserved (Fig 70) The P wave is almost always easily discernible although occasionally it may be superimposed on the preceding T wave Sinus tachycardia may be the

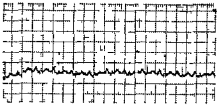


Fig 70

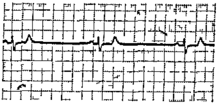


Fig 71

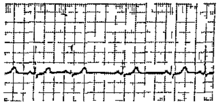


Fig 72

Figure 70 Sinus tachycardia
Figure 71 Sinus bradycardia
Figure 72 Sinus arrhythmia

result of physiologic conditions (excitement exercise) pathologic situations not necessarily involving the heart (fever) and finally may be the first expression of cardiac embarrassment (carditis congestive failure) Reflex vagal stimulation (carotid sinus pressure) results only in a gradual decrease of the rate P waves previously superimposed on the T waves may thus be clearly visible

Sinus bradycardia (Fig 71) with rates of less than 100 beats per minute in infants and less than 80 in children is usually an expression of good cardiac reserve Occasionally however it may be seen in cases of rheumatic carditis Exercise should raise the rate in sinus bradycardia with ease in contrast to the relatively fixed rate of complete atrioventricular block

Sinus arrhythmia (Fig 72) is a phasic irregularity of the heart beat speeding up with inspiration and slowing with expiration but maintaining the conventional P QRS relationship Much like sinus bradycardia it may be an expression of good cardiac function it is commonly seen in older children and adolescents seldom in young children and almost never in infants Slower rates predispose to it Sinus arrhythmia is frequently present during convalescence

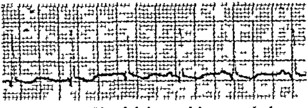


Figure 73 Sudden shift from nodal to sinus rhythm

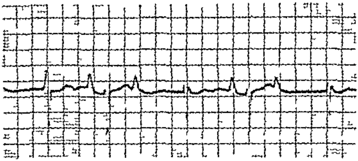


Figure 74 Sinoatrial block The second P wave in the tracing is blocked not followed by a QRS complex Similarly the fourth one is blocked as well

A wandering pacemaker representing a gradual shift through several cycles from the sinoatrial to the atrioventricular node is pictured in the electrocardiogram by a change from a normal upright P wave with an average P R interval to an abnormal often inverted P wave with a short even negative P R interval A shifting pacemaker is a sudden jump from sinus rhythm to nodal pacemaker (Fig 73) Once the atrioventricular node becomes the pacemaker the process reverses itself and the impulse formation gradually reverts to the sinoatrial node This phenomenon is commonly seen in people without heart disease but it does occur with rheumatic fever or digitalis intoxication

Sinoatrial block and sinus arrest are temporary suspensions of the electrical activity of the atria through two or more cycles (Fig 74) Sinoatrial block is usually episodic and of relatively short duration the sinus pauses are multiples of the normal sinus rhythm Sinus arrest is of longer duration and the length of the pauses bears no mathematical relation to the original cycle length Both conditions may be secondary to increased vagal tone anoxia or drugs (digitalis quinidine)

Rhythms Originating in the Atrium or the Atrioventricular Node These are called supraventricular rhythms In contrast to the sinus rhythms either no P waves are noted or the atrial waves are unusual in contour and are separated from the QRS complex by less than 0 10 second With few exceptions the QRS complexes of supraventricular rhythms are of normal configuration and are less than 0 10 second in duration

The least significant of the supraventricular arrhythmias is the supra

Sinu ta hy rdia

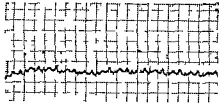


Fig 70

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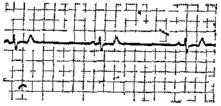


Fig 71

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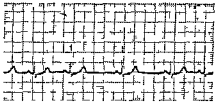


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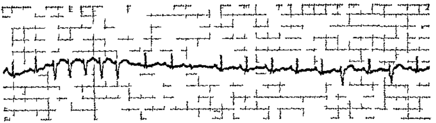


Figure 77 Repetitive tachycardia Note the occurrence of ectopic beats in short bursts and then singularly

premature beats are common in healthy youngsters even in newborn infants and may have no significance whatsoever They may also accompany structural heart disease but the diagnosis must rest on additional evidence Supraventricular premature beats are characterized by a less than complete compensatory pause that is the length of two cycles including one with the premature beat is less than the length of two normal cycles

A continuous series of supraventricular premature beats ranging in duration from a few seconds to several years is called paroxysmal atrial tachycardia (Fig 76) This then represents a complete temporary usurpation of the function of the sinoatrial node by an atrial or nodal pacemaker The rate may vary from 160 to 300 beats per minute The onset as well as the termination is sudden in most instances The rate is completely fixed and unchanging within a fraction of a second Reflex vagal stimulation either results in the re establishment of sinus rhythm or has no effect at all P waves are seldom if ever seen T waves are usually inverted if the tachycardia has lasted for more than a day Paroxysmal supraventricular tachycardia often occurs in a child with a normal heart nevertheless if it lasts long enough it may cause congestive failure and even death Obviously this is even more likely to happen if the patient has underlying heart disease

A transition between paroxysmal atrial tachycardia and supraventricular premature beat is the so called repetitive tachycardia [68] in which short bursts of paroxysmal tachycardia alternate with single premature beats from the same focus (Fig 77) This is usually seen in young people and infants with otherwise normal hearts

One of the severest supraventricular arrhythmias is atrial flutter The atria here are in continuous regular activity ranging from 200 to 350 beats per minute The ventricles respond with normal QRS complexes in a regular fashion (1 1 to 1 4 ratio) at regular rates of 140 to 160 beats per minute Characteristically in lead I the flutter waves can hardly be noted whereas in leads II and III they clearly appear in triangular form (Figs 49 78) This is a severe arrhythmia and almost always means organic heart disease specially rheumatic carditis Occasionally it may be encountered in children with congenital heart disease

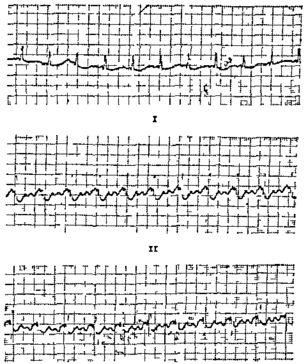


Figure 78 Atrial flutter

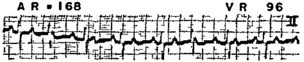


Figure 79 Paroxysmal tachycardia with block Note the moderately fast atrial rate with a prolonged P R interval and an occasionally blocked atrial wave such as no 6

or subendocardial sclerosis and it may even occur in normal hearts as paroxysmal supraventricular tachycardia The effect of atrial flutter on the circulation as a whole depends on the ventricular rate As long as the ventricles do not respond to more than one third of the atrial impulses there usually is little difficulty If the ventricular response is 1 2 or even 1 1 congestive failure will ensue Reflex vagal stimulation can increase the atrioventricular block and reduce the ratio of ventricular response from 1 2 to 1 3 or even 1 4 often this is the only way by which clear flutter waves unobstructed by QRS complexes can be identified

An arrhythmia encompassing certain features of atrial flutter and certain characteristics of supraventricular tachycardia is the one called paroxysmal atrial tachycardia with block (Fig 79) [3] In this arrhythmia the atria beat at a moderately fast rate 140 to 180 beats per minute and the ventricles respond irregularly in 1 1 to 1 3 fashion The P R

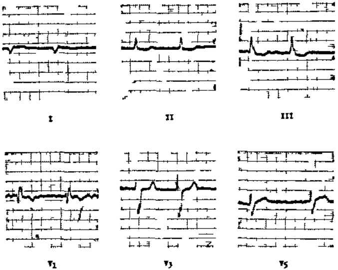
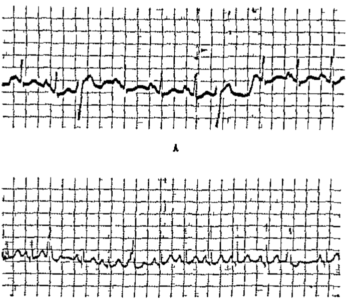


Figure 80 Atrial fibrillation

interval is unusually long (0 14 to 0 20 second) and an occasional P wave is completely blocked

The severest type of supraventricular arrhythmia is atrial fibrillation (Figs 50 80) This is a continuous irregular activity of the atria at rates of 400 to 700 beats per minute The atrial activity may manifest itself electrocardiographically only as a minor irregularity of the base line so called fibrillary wave at other times the atrial waves are coarse and relatively slower closely resembling flutter waves These various types of fibrillatory activities may be seen at different times in one and the same tracing The ventricles respond irregularly but at a much reduced rate (110 to 150 beats per minute) Both the atrial and the ventricular beats are totally irregular Atrial fibrillation for practical purposes always means severe organic heart disease It is most common in rheumatic heart disease but can be seen in congenital heart disease especially atrial septal defect Ebstein s disease or tricuspid atresia The effect of atrial fibrillation on the circulation depends for the most part on the ventricular rate If this is relatively slow atrial fibrillation may be *tolerated for a long time without symptoms On the other hand* unusually fast ventricular responses often lead to catastrophe That atrial fibrillation even with a slow ventricular rate does not result in optimal circulatory dynamics is demonstrated by the fact that conversion of these slowly beating fibrillatory rhythms to normal sinus rhythm with the same ventricular rate is accompanied by great subjective improvement of the patient Reflex vagal stimulation if effective at all results in gradual slowing of the ventricular rate

Rhythms Originating in the Ventricle These rhythms are characterized by a lack of correlation between atrial and ventricular complexes



A

B

Figure 81 Ventricular premature beats lead I *A* Origin of the premature beat in the left ventricle *B* in the right ventricle

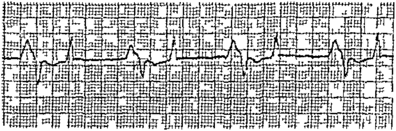


Figure 82 Bigeminy Ventricular premature beats alternating with supraventricular beats

The P waves are normal in shape The ventricular complexes on the other hand are bizarre in configuration and measure 0 10 second or more the QRS and T waves often point in opposite directions Lead I usually gives the clue as to whether the ventricular complex originates from a focus in the right or the left ventricle (Fig 81) Right ventricular beats resemble left bundle branch block complexes and have a dominant R in lead I whereas left ventricular beats similar to right bundle branch block show a deep S_1

Ventricular premature beats are the most innocuous among the ventricular arrhythmias Their significance in pediatrics is only slightly greater than that of supraventricular premature beats They may occur in perfectly normal youngsters on the other hand they may indicate carditis anoxia or digitalis or quinidine intoxication Ventricular premature beats alternating with normal beats so called bigeminy (Fig

I



Figure 83 Burst of ventricular tachycardia after two beats with normal sinus rhythm After the tachycardia the sinus rhythm recurs and then is followed by another premature beat

82) are especially characteristic of digitoxicity The occurrence of multifocal multiple shaped ventricular premature beats has a more serious significance than if the shape of the extrasystoles is always the same within one lead The characteristic feature of ventricular premature *beats in addition to their shape and their lack of time correlation with* atrial activity is the fact that they are followed by a full compensatory pause This means that the length of two cycles including one premature beat is the exact length of two normal cycles

A series of ventricular premature beats at a rate of 120 to 180 per minute constitutes ventricular tachycardia (Fig 83) This phenomenon is rare in children so much so that when it does seem to occur suspicion is immediately aroused as to whether it really represents ventricular tachycardia or supraventricular tachycardia with aberrant intraventricular conduction

The differentiation between these two conditions is sometimes difficult but usually is possible on the basis of the following points (1) Supraventricular tachycardia is influenced by reflex vagal stimulation (carotid sinus or sinus of Valsalva) ventricular tachycardia is not (2) The *P* waves in supraventricular tachycardia always show some regularity in relation to the QRS complex whereas no such connection exists in true ventricular tachycardia (3) The rates of the P and QRS complexes are similar in supraventricular tachycardia whereas they are totally different in ventricular tachycardia (4) The rate of supraventricular tachycardia is characterized by a clocklike regularity not present in true ventricular tachycardia

The dangers of ventricular tachycardia are twofold It may result in congestive failure if the rate is rapid enough and lasts long enough or it may progress into ventricular flutter or fibrillation The latter situation is not compatible with effective cardiac output thus it results in sudden death Because of this possibility of conversion into ventricular flutter *or fibrillation paroxysmal ventricular tachycardia is regarded much more seriously* than is paroxysmal supraventricular tachycardia

Ventricular flutter and fibrillation are terminal events and recovery from them is relatively rare They represent the electrocardiographic substrate of sudden death The complexes are broad slurred and notched and the T waves are fused with the QRS complexes The amplitude varies tremendously within one strip Many more tracings of

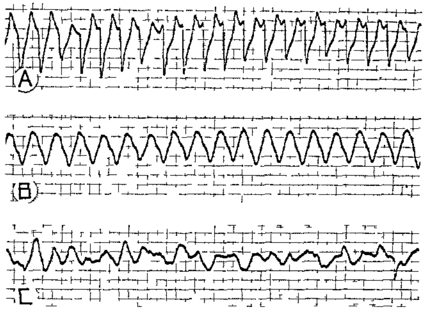


Figure 84 A Ventricular tachycardia giving rise in (*B*) to ventricular flutter and in (*C*) to ventricular fibrillation These are consecutive strips of a patient who died after cardiac surgery

this sort have been obtained during cardiac operations within the past ten years than were seen in the entire previous history of electrocardiography (Fig 84)

Effects of Chemical Changes and Drugs on the Electrocardiogram

Potassium The effect of potassium (Fig 85) on the electrocardiogram has been amply documented both in animal experimentation and in man [30, 110] Whether the electrocardiogram is principally influenced by the potassium level in the serum or within the myocardium itself is the subject of considerable argument It seems however that the proponents of the cellular theory have made a better case

Potassium intoxication has been produced in animals by infusion or ingestion of large amounts of potassium salts In man potassium poisoning is encountered in the later stages of uremia leukemia and lower nephron nephrosis and after treatment with excessive amounts of potassium The heart rate usually slows and the P waves become progressively wider and finally disappear while the P R interval gradually increases The QRS complex undergoes changes consisting in widening and diminution of the R waves and increase of the S waves The T waves become taller and tent shaped and eventually surpass the R wave in height (Fig 58) No definite correlation exists between serum potassium level and electrocardiographic changes beyond the observation that changes are rarely missed if the serum potassium level is 7 mEq per liter or more unless the level of sodium in the blood is unusually low

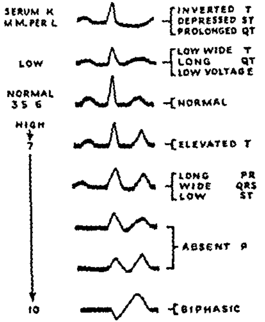


Figure 85 Schematic representation of the changes in P QRS T complex in conjunction with potassium levels (From Darrow, D. C. New England J. Med. Vol. 242.)

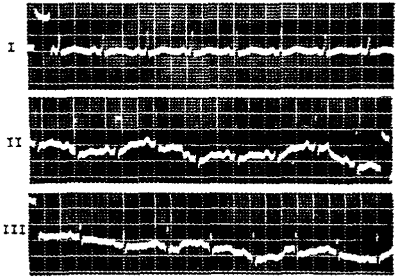


Figure 86 Potassium depletion in the electrocardiogram

Potassium depletion seen in man in postoperative alkalosis, diabetic coma, long standing diarrhea and vomiting, also causes typical changes (Fig. 86) consisting in a prolongation of the Q T interval with widening of the T waves and occasional depression of the S T segment.

Calcium Calcium depletion, particularly in cases of tetany also

I

II

III

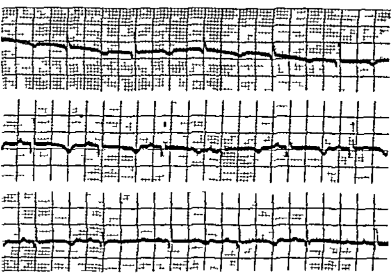


Figure 87 Calcium depletion

causes prolongation of the Q T interval (Fig 87)[30, 110] In contrast to potassium depletion however the lengthening of the electrical systole is caused by a prolongation of the S T segment rather than by a widening of the T wave the T wave in hypocalcemia is relatively normal perhaps slightly narrow The tetanic contraction of the striated muscle is reflected in the somatic tremor noted on the limb leads

A high level of serum calcium hypercalcemia may cause increased irritability of the heart with premature beats even paroxysmal ventricular tachycardia [95] It is also said to shorten the Q T interval

Digitalis The electrocardiogram may be used to determine whether significant amounts of digitalis have been given to a patient and with some qualifications whether toxicity is present The electrocardiogram cannot answer the question Has the patient been fully digitalized? Whether or not full digitalization has been achieved is a question for the clinician to decide the electrocardiogram only helps to determine whether there is some or too much digitalis in the heart muscle

The digitalis glycoside in doses commonly given does not influence the shape of P waves However the development of prolonged atrioventricular conduction time or sinoatrial block may be considered a sign of digitoxicity (Fig 88) There are no significant changes in the QRS complex with doses usually given in man Changes in the S T segment are comparatively common by changing the ventricular gradient S T segments hitherto isoelectric or elevated are depressed and those that have been depressed are raised by the drug (Fig 89) The S T changes are most obvious in leads with a tall R wave and consist in a straight angular "step down" appearance of the S T segment S T changes caused by digitalis administration often cannot be distinguished from those

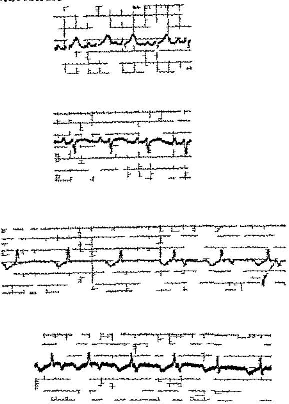


Figure 88 Consecutive daily tracings in patients with digitoxicity The top tracing shows normal sinus rhythm The second tracing shows prolonged P R interval The third tracing shows changing pacemaker and the fourth one show a return of this changing pacemaker to sinus rhythm with progressively prolonged P R interval

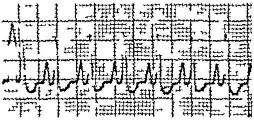
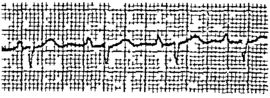


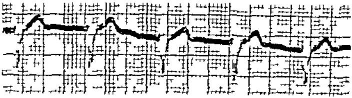
Figure 89 Typical S T depression in patient with digitalis intoxication

resulting from myocardial damage however the straightness and the angularity of the depression without any convexity favor a digitalis effect Digitalis affects the T waves in much the same way it affects the S T segments positive T waves become inverted and negative T waves become upright A shortening of the Q T interval in my experience is the most consistent effect of digitalis on the electrocardiogram

One of the earliest and most favorable results of digitalis administration is a slowing of the heart rate seldom if ever does one see a



A



B

Figure 90 The effect of quinidine on the electrocardiogram A Normal sinus rhythm B Nodal rhythm with widening of the QRS complex

favorable digitalis effect without a significant drop in rate This probably represents increased vagotonia and develops as a result of sinus depres sion in cases of sinus rhythm and because of increasing atrioventricular block in cases of atrial flutter or fibrillation Undue slowing of the rate to less than 100 beats per minute in infants and less than 80 in older children is presumptive evidence of digitoxicity

The development of arrhythmias in most instances is evidence of digitoxicity the one exception is the appearance of a sinus arrhythmia which may simply be the concomitant of the slower rate and increased vagotonia On the whole digitalis intoxication results more commonly in atrial arrhythmias in children than in adults in whom ventricular arrhythmias seem to dominate the picture Ectopic atrial pacemakers and paroxysmal atrial tachycardias with block are seen relatively fre quently as a first expression of digitoxicity Atrial flutter and fibrillation may also be seen although much less commonly and they represent a much more serious degree of intoxication Ventricular arrhythmias are not seen frequently in children Ventricular premature beats if present are prone to develop in forms of bigeminy

If a patient is receiving digitalis and an arrhythmia develops the burden of the proof is on the physician who maintains that the change in rhythm whatever its nature was not caused by digitalis The diffi culties resulting from this statement are obvious sometimes it may be extremely hard to decide whether a patient has received too much or too little digitalis The only reasonable approach to this problem is to try to withhold the drug for a few days if possible and to observe the trend of the changes If the patient is in severe congestive failure so that withholding the glycoside appears dangerous then administration of small frequent doses of a rapidly excreted form of digitalis is recom mended with electrocardiographic control [1]

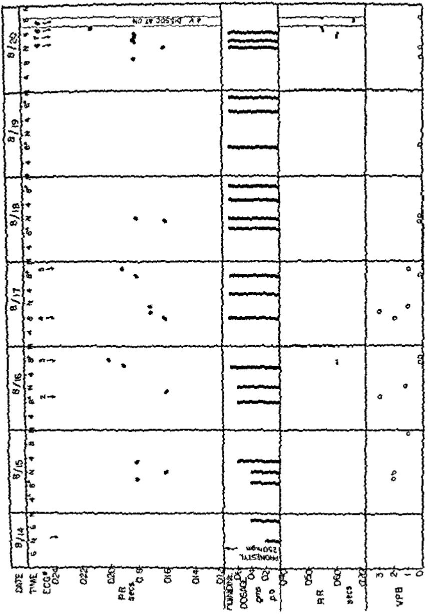


Figure 91 The effect of quinidine on the P R interval This is a schematic presentation of the changes (From J Merkes A S Nad and A Linenthal)

The time relationship of digitalis administration on the electrocardiogram must be mentioned briefly In my experience the earliest effects are the shortening of the Q T interval and the changes in the T waves The rapidity of the changes and their duration depend a great deal on the preparation used Digitalis even in relatively large doses affects normal hearts in a minor fashion only if at all

Quinidine The effects of quinidine on the electrocardiogram consist in slowing of the pacemaker and widening of the QRS complex the Q T interval and the T waves (Fig 90)

It has been my practice to consider a widening of the QRS complex by more than 50 per cent as evidence of definite toxicity Other toxic effects include the appearance of multifocal ventricular premature beats and prolongation of the P R interval (Fig 91)

Electrocardiographic changes after a single oral dose of quinidine appear to be maximal in about one hour and they practically disappear in a matter of four hours

Pronestyl The effects of procaine amide are essentially similar to those seen with quinidine administration doses causing a 50 per cent prolongation of the QRS complex are considered toxic Occasionally supraventricular premature beats with aberrant conduction ventricular tachycardia and even ventricular flutter have been seen as a result of Pronestyl toxicity

Pericarditis

The electrocardiographic features noted in patients with pericarditis may be the result of two factors One is pericardial fluid accumula

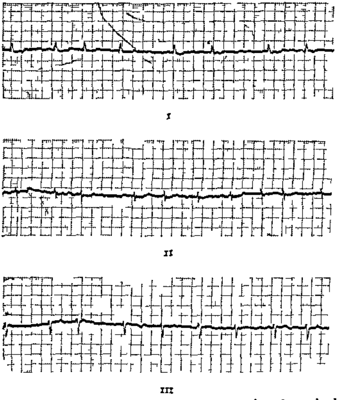


Figure 92 Tracing in a patient with constrictive pericarditis Note the low voltage QRS complexes throughout and the presence of atrial fibrillation

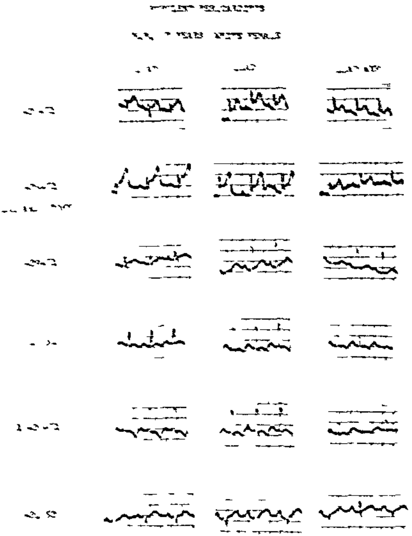


Figure 93. Progressive changes in the electrocardiogram of a patient with purulent pericarditis. Note the appearance of deep S-T and T wave inversions a considerable period after pericardiotomy and clinical cure of the patient. Note the marked S-T elevation before operation.

tion, or thick scarring (constrictive pericarditis) which results in low voltages throughout all leads (Fig. 92). This feature is neither distinctive nor constant; it may be seen in other instances of fluid accumulation such as the pleural effusion of nephrosis, and it may not be present in severe and advanced cases of pericarditis.

The second type of change—resulting from subepicardial myocardial damage—is more constant and more characteristic. It consists in a concave elevation of the S-T segments and T waves throughout all leads, at first. Later the S-T segments return toward normal and the T waves

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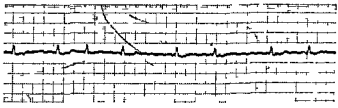
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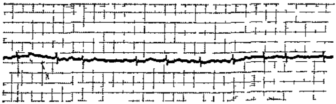
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PERICARDITIS

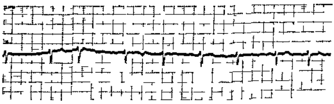
The electrocardiographic features noted in patients with pericarditis may be the result of two factors One is pericardial fluid accumula



I



II



III

Figure 92 Tracing in a patient with constrictive pericarditis Note the low voltage QRS complexes throughout and the presence of atrial fibrillation

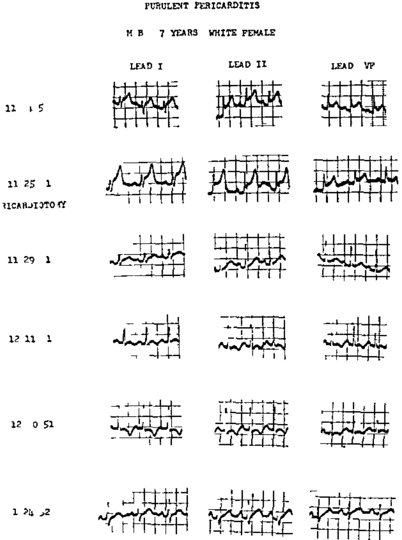


Figure 93 Progressive changes in the electrocardiogram of a patient with purulent pericarditis Note the appearance of deep S T and T wave inversions a considerable period after pericardiotomy and clinical cure of the patient Note the marked S T elevation before operation

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The second type of change—resulting from subepicardial myocardial damage—is more constant and more characteristic It consists in a concave elevation of the S T *segments* and T waves throughout all leads at first Later the S T segments return toward normal and the T waves

become negative Finally sharp inversion of the T waves is present with perfectly normal S T segments (Fig 93) The time relationship of these changes is interesting The S T elevation occurs early usually within a week of the appearance of a friction rub The sharp T wave inversion occurs late (three to four weeks after the onset of the disease) often at a time when all clinical evidences of pericarditis have disappeared Severe T wave changes may persist one to two months after complete clinical recovery has taken place Thus during the time of the most severe clinical changes the electrocardiogram may be perfectly normal *the S T segments have already returned to normal and the T wave* inversions have not yet become significant

A differential point of almost no importance in pediatrics should be mentioned briefly The S T and T wave changes of pericarditis are similar to those seen in myocardial infarction with two exceptions (1) The changes observed in pericarditis are not usually localized to any one area in the chest leads (2) In pericarditis the changes in the S T segment are usually absent by the time inversion of the T waves occurs, whereas in myocardial infarction the two changes are more or less synchronous

P wave changes of the "P mitrale" type are commonly seen in constrictive pericarditis

Phonocardiography

USE AND EQUIPMENT

In view of the emphasis placed on auscultation in the diagnosis of heart disease in children it seems logical to devote a small section of this book to phonocardiography the graphic registration of heart sounds This is an excellent tool for the teaching of intelligent interpretation of auscultatory phenomena It also serves the useful purpose of enabling one to check the accuracy of ones own auscultation and occasionally plays the role of an unbiased arbiter deciding between two contrary opinions in regard to murmurs and sounds In this latter respect it is especially helpful for the younger physician or the medical student who with his own accurate observations now reinforced by the phonocardiogram can often successfully challenge his elders and teachers Phonocardiography may thus prove to be a revolutionary instrument in medicine substituting facts for authority derived from the gray hair of the teacher enhanced by his academic standing but sadly limited by the gradually narrowing hearing acuity of age

Excellent monographs and papers on phonocardiography [07, 2, 4, 30, 6, 78, 79] are available in the literature hence no complete review of the subject will be attempted here

The only phonocardiograph with which I am well acquainted is the Sanborn Twin Beam In this recorder two simultaneous tracings are obtained The top tracing represents the heart sounds and murmurs the bottom one is used as a reference tracing necessary for timing The electrocardiogram is used most frequently for reference but for certain purposes registration of the jugular venous pulse the carotid pulse or the apex beat (linear or apex cardiogram) is more suitable

Three kinds of microphones are available with this apparatus The so called linear microphone is sensitive to all the vibrations caused by the heart and as such registers primarily the coarse vibrations (less than 100 per second) set up by the cardiac impulse (Fig 94 *A*) the jugular vein or the carotid artery and suppresses the finer higher frequency vibrations constituting the sounds and murmurs

The stethoscopic microphone on the other hand completely suppresses the coarsest elements and registers only the low frequency sounds some of them inaudible (between 30 and 200 per second) offered to the ear by the stethoscope (Fig 94 *B*) The higher frequency

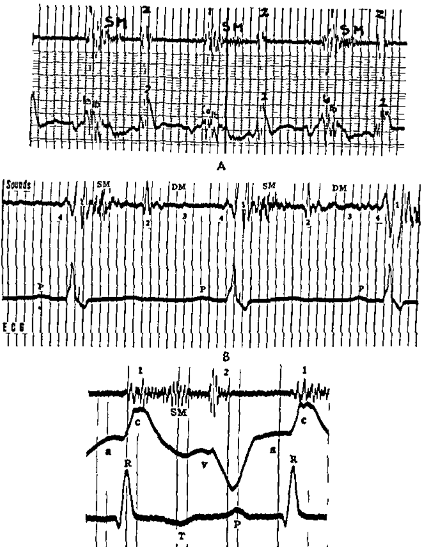


Figure 94 A Upper line logarithmic phonocardiogram Lower tracing linear cardiogram At the upper tracing note *1* and *2* indicating first and second heart sounds and *SM* indicating systolic murmur Lower tracing *1a 1b* and *2* are waves of the linear cardiogram For schematic representation of the components of the linear cardiogram (apex cardiogram) see Figure 95

B Upper tracing stethoscopic phonocardiogram Lower tracing electrocardiogram Numbers *1 2 3 4* indicate first second third and fourth heart sounds *SM* means systolic murmur *DM* equals diastolic murmur

C Three simultaneous tracings Upper line logarithmic phonocardiogram Middle line jugular venous pulse Lower line electrocardiogram The phonocardiogram and the electrocardiogram are marked as in (*A*) and (*B*) The jugular venous pulse is marked according to the conventional markings of jugular venous pulse See Figure 95

murmurs are not brought out well in the stethoscopic tracing The use of this microphone is principally in the analysis of the heart sounds it is of less use in demonstrating murmurs

The logarithmic microphone has the same kind of "bias" as the human auditory cortex It suppresses not only the lowest vibrations of the cardiac impulse but also the low frequency murmurs and sounds and brings out with clarity and logarithmic intensity the highest fre quency vibrations (50 to 500 per second) (Fig 94 C) The logarithmic tracing forms the basis of most of our phonocardiographic studies it is especially useful in the analysis of murmurs

For the majority of the clinical problems sound tracings obtained by the logarithmic microphone with electrocardiographic reference are entirely satisfactory For closer analysis of the heart sounds a stetho scopic and a logarithmic tracing obtained from the apex with jugular venous linear (apex) cardiographic and carotid arterial reference tracings is desirable

Three types of chest pieces are used commonly with the Sanborn Twin Beam—a medium open bell a small open bell and a Bowles diaphragm The first two are used for registering heart sounds and low frequency murmurs the third is more advantageous for high frequency insufficiency blows and continuous murmurs

On the basis of phonocardiograms taken with the instrument, mi crophone and chest pieces just discussed rather accurate analysis of the auscultatory phenomena observed over the hearts of children is possible

A direct writing phonocardiograph with six filters has been used to good advantage by Mannheimer and his associates [191 36] Unfortunately I have had no personal experience with this apparatus hence the reader seeking information on this particular machine is referred to the work of the Swedish authors

HOW TO TAKE A PHONOCARDIOGRAM

A quiet room is a prerequisite to the taking of a clear phonocardio gram Soundproofing is useful but not essential Telephones and type writers should not be used at the time a tracing is taken

The child should rest comfortably on an examining table or should be propped up on a pillow in bed Only under extremely fortunate circumstances may satisfactory tracings be obtained from children less than five years old without heavy sedation The cooperation of an older child may easily be enlisted even to the extent of having him hold his breath if the procedure is explained to him

At The Children s Hospital in Boston it has been our custom to obtain logarithmic tracings with an open bell chest piece from the apex the left sternal border and the second left and second right interspaces for all patients using the electrocardiogram as a reference We also

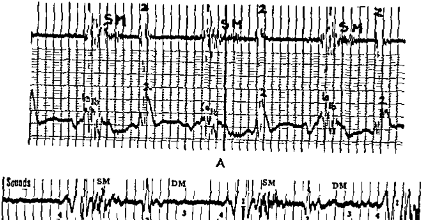


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take logarithmic and stethoscopic tracings from the apex with the linear cardiogram and the jugular venous pulse as a reference tracing In addition to these routine points of auscultation tracings may be obtained from any other location desired All these should be taken in a definite consistent sequence so that when the tracing is developed and analyzed the identity of each strip will be clear purely on the basis of its position within the series It is also important as a further aid in identification that a permanent notebook be kept containing the patient s name record number age diagnosis and particularly the sequences of each tracing One should also note the type of microphone the chest piece the amplification and the nature of the reference tracing used for every location

HEART SOUNDS

Four heart sounds are normally registered on a stethoscopic tracing (Fig 95) The first sound or sound complex is approximately synchronous with the QRS complex in the electrocardiogram its average duration is 0 10 second (0 05 to 0 15) Its onset can be well timed by

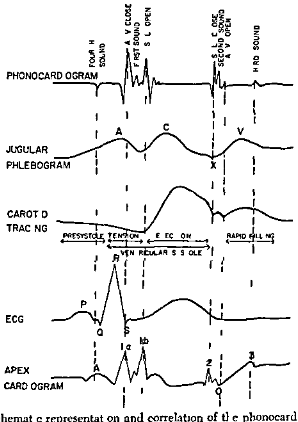


Figure 95 Schemat c representat on and correlation of tl e phonocard ogram j g l r phlebogram carot d tracing electrocard ogram and apex card ogram

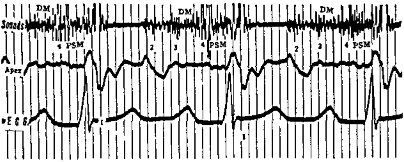


Figure 96 Late first sound

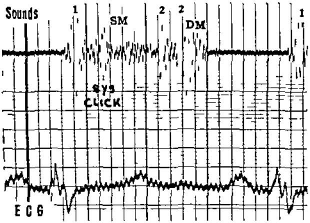


Figure 97 A systolic click in patient with pulmonary arterial hypertension and dilated main pulmonary artery

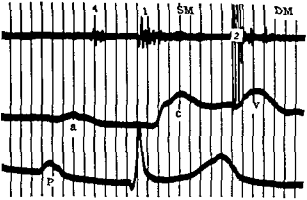


Figure 98 Upper tracing phonocardiogram Middle tracing jugular venous pulse Lower tracing electrocardiogram Note the low intensity first sound in the phonocardiogram

the q wave but its termination cannot be timed electrocardiographically the C wave of the jugular venous pulse probably serves best as an end point The main portion of the first sound results from the closure of the mitral and tricuspid valves with some addition from the semilunar opening and atrial and vascular components

Abnormalities of the first sound can be well documented phono cardiographically An accentuated and occasionally late first sound (Fig 96) is characteristic of mitral stenosis large ventricular septal defect or pulmonary stenosis Levine[97] pointed to the presence of a loud first sound in patients with a short P R interval A systolic click occurring after the semilunar valves have opened may be heard at the second right interspace in patients with aortic stenosis[91] and at the second left inter space in patients with pulmonary stenosis[9] or pulmonary valve obstruction (Fig 97) A low intensity first sound is observed in patients with pericardial effusion myocardial failure or prolonged P R interval (Fig 98)

The second sound or sound complex although usually occurring beyond the T wave does not show a close correlation with the electro cardiogram More accurate timing is possible by using the linear or apex cardiogram as a reference tracing The second sound begins with the 2 wave *and ends with the 0 point (Fig 95) Its average duration is 0 08* second (0 03 to 0 12)

The closure of the semilunar valves contributes the major portion of the second sound with some muscular and vascular elements added The degree of splitting between the semilunar elements of the second sound should be carefully noted The two sets of semilunar valves—aortic and pulmonary—close practically simultaneously in the normal adult the vibrations caused by the closure of these valves are thus super imposed on each other hence no splitting occurs In contrast in children under normal circumstances the semilunar valves close asynchron ously[5] the aortic first and the pulmonary second Consequently the trained ear may easily discover a slight splitting accentuated on deep inspiration within the second sound of normal children If measured phonocardiographically the two peaks constituting the splitting normally are seldom more than 0 04 second apart

One other component of the second sound is the so called opening sound of the mitral valve This is the terminal portion of the second sound complex is identified by the 0 point of the linear cardiogram and consists normally of vibrations too low to be audible or visible in the logarithmic tracing However it can be seen well in the stethoscopic tracing obtained from the apex

The abnormally high amplitude second sound at the second left interspace is typical of patients with pulmonary arterial hypertension[94] (Fig 99) In contrast the second sound in patients with pulmonary stenosis is of low intensity at the pulmonary area[9] (Fig 106 *B*) Both

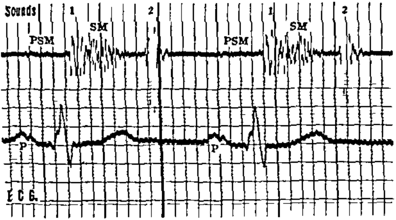


Figure 99 Upper tracing logarithmic phonocardiogram Lower tracing electrocardiogram Note the high amplitude second sound in the phonocardiogram

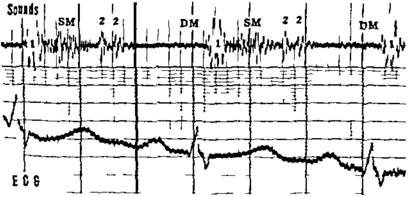


Figure 100 Widely split second sound Upper tracing logarithmic phonocardiogram Lower tracing electrocardiogram

these conditions affect only the second or pulmonary element of the semilunar component Wide splitting (exceeding 0 05 second) of the second sound at the second left interspace with moderately intense aortic and pulmonary elements is typical of atrial septal defect [5] (Fig 100) Wide splitting with a low intensity pulmonary element is characteristic of severe cases of pulmonary stenosis The opening sound of the mitral valve normally present only in the stethoscopic tracing is clearly demonstrable as an opening snap (Fig 101) in logarithmic tracings of many patients with mitral stenosis [61]

The third heart sound is identifiable by the 3 wave in the linear cardiogram (Fig 95) and corresponds to the first rapid inflow of blood from the atria to the ventricles Usually this is clearly audible in children less commonly in adults It is well registered in the logarithmic as well as in the stethoscopic tracing The fourth heart sound or atrial sound

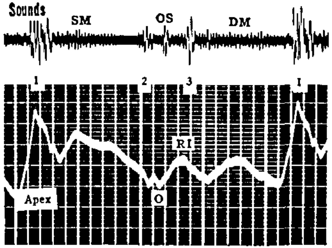


Figure 101 Opening snap in patient with mitral stenosis Upper tracing phonocardiogram lower tracing apex or linear cardiogram Note O at opening snap corresponding to O point in the linear cardiogram (From Ongley P A Sprague H B and Rappaport, M B New England J Med Vol 253)

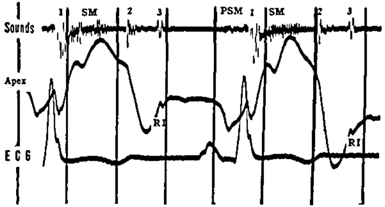


Figure 102 Gallop rhythm consisting of first sound second sound and loud third sound Upper tracing logarithmic phonocardiogram Middle tracing apex cardiogram Lower tracing electrocardiogram

corresponds to the second rapid filling period of the ventricles and is identifiable by the fact that it follows the P wave and precedes the q wave in the electrocardiogram (Fig 95) This sound consists of low frequency vibrations and is usually not heard in normal people but is well discernible in a stethoscopic phonocardiogram

Unusually high amplitude or high frequency third (protodiastolic) and fourth (presystolic) heart sounds with tachycardia are recognized with a stethoscope as gallop rhythm (Fig 102) In most instances this means cardiac failure A well registered third sound in an adult may be the precursor of the mid diastolic murmur of mitral stenosis A high amplitude fourth or atrial sound is commonly registered in patients

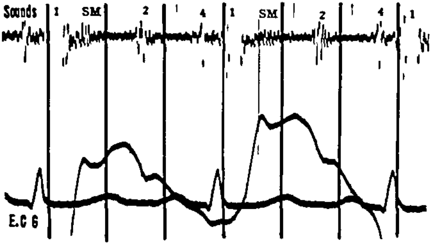


Figure 103 Loud atrial or fourth sound in logarithmic tracing Upper line logarithmic phonocardiogram Middle line jugular venous pulse Lower line electrocardiogram

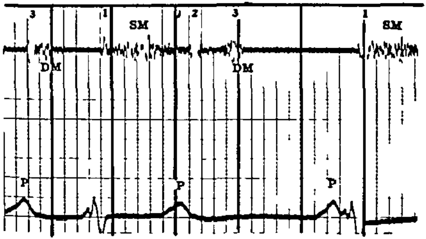


Figure 104 Phonocardiogram of complete heart block. Note the irregular relationship between P wave and QRS complex in the electrocardiogram and the variable intensity of the first heart sound depending on the vicinity of the P wave and the QRS complex The first sound after the first QRS complex is low in intensity since the P R interval here is long The first sound after the second QRS complex is high in intensity and P R interval short.

with right atrial hypertrophy (Fig 103) (severe pulmonary stenosis tricuspid atresia total pulmonary venous anomaly and Ebstein s disease) and in patients with complete heart block (Fig 104)

MURMURS

Vibrations with frequencies of 50 to 500 per second registered between heart sounds are called murmurs They are thought to result from turbulences developing in the laminar flow of blood Occasionally

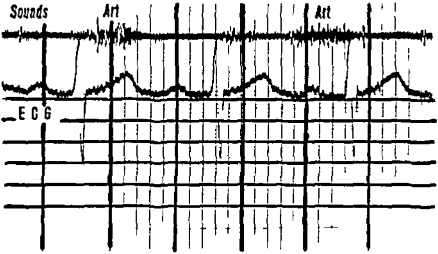


Figure 105 Respiratory artefact

it may be difficult to distinguish a short murmur from a prolonged heart sound In most instances however murmurs have a duration long enough to make differentiation relatively easy

Occasionally a more serious problem is presented by the occurrence of artefacts (respiratory sounds noises in the room defects in the ma chinery and so forth) The best way to avoid these is to be scrupulously certain that the phonocardiogram is taken in a quiet room and that the base line between sounds is kept as straight as possible Furthermore one ought to suspect all vibrations extending with equal intensity and frequency throughout systole and diastole especially if appearing peri odically rather than constantly of being artefacts of respiratory origin (Fig 105)

Once the vibrations of appropriate frequency and timing have been identified in the phonocardiogram as murmurs their significance has to be decided upon This brings one to the question of organic versus functional murmur a terminology I have tended to discard in favor of significant and not significant" murmur The auscultatory phenomena associated with permanent anatomic deformities of the heart are com monly called organic murmurs and have fairly severe implications Logically one would include in this group conditions with such varying significance as mitral stenosis the mildest form of pulmonary stenosis aortic regurgitation and small Roger type ventricular septal defect By contrast in the functional category with a usually benign connotation would be included conditions without permanent abnormalities such as anemia rheumatic carditis without valvular deformities fever or thyro toxicosis

This differentiation based on pathologic data is meaningless today in view of the physiologic information available It may readily be seen

that mild pulmonary stenosis with a gradient between the right ventricle and the pulmonary artery of 15 to 20 mm of mercury giving rise to a loud systolic murmur may never be demonstrable at postmortem examination Will such a murmur be called "organic" or "functional? Conversely shall we classify as "organic" or "functional" the loud murmur of a small ventricular septal defect without demonstrable left to-right shunt? The terms "organic" and "functional are clearly wrong in their prognostic implications A small ventricular defect though "organic" may be compatible with a long and active life whereas the "functional" murmur of severe anemia may be associated with severe cardiac failure

Finally if one reviews the available literature to learn which murmurs most authors call "functional" and which "organic" one concludes *that all loud systolic murmurs and everything that occurs in diastole is* called "organic" whereas everything that is faint in systole is called "functional" The fallacy of this type of reasoning has been proved repeatedly by the finding of a loud early diastolic murmur in the pulmonary area of a patient with an anatomically normal heart (idiopathic pulmonary regurgitation) and by the presence of only a faint systolic murmur in patients with pulmonary atresia or the severest type of mitral stenosis

In view of these difficulties then the murmurs should be properly described in respect to timing intensity duration and character and then should be correlated with the other necessary and available clinical and laboratory information and designated as being "significant" or "not significant at that particular time

*Murmurs associated with cardiac symptoms x ray evidences of car*diac enlargement or abnormal electrocardiograms should certainly be considered "significant" Even without these ancillary findings a murmur is likely to be "significant" if it is of high intensity (more than grade 3 according to Levine) and appreciable duration if it transmits well throughout the precordium the neck and the back and if it is not influenced by the phases of respiration or change in position Diastolic murmurs on the whole are more likely to be "significant" than systolic ones

The "insignificant" murmur is never accompanied by cardiac symptoms and laboratory evidences of heart disease Usually it is a murmur of low intensity low frequency and of short duration occupying mid systole It may be heard maximally at the left sternal border or the apex it transmits poorly and it shows considerable variations from cycle to cycle with respiration change in position or for no very obvious reasons A particular type of "insignificant murmur commonly heard in healthy *adolescents is strongly localized to the left lower sternal border* and sounds much like the plucking of a string

A word may be said here about the occurrence character and significance of murmurs in children Richards [39] reporting on more than

5000 newborns found a systolic murmur at birth in 1 7 per cent a figure in close agreement with that published several years ago by Lyon and his associates [30] These studies confirm the impression of most pediatricians that murmurs in the newborn are relatively rare The character of these murmurs varies a great deal I have the distinct impression that some of them are rather harsh even "stenotic" although the majority are obviously soft and blowing It is commonly stated that murmurs in the neonatal period may originate from a patent foramen ovale or a patent ductus arteriosus Though the validity of such a contention cannot be categorically denied, attention should be directed to two facts (1) Patients with patent ductus arteriosus never have continuous murmurs in infancy (2) Children with proved patency of the foramen ovale almost never have murmurs attributable to a shunt through this defect My own view of the matter is that we do not know where the occasional murmur in the newborn originates and it almost seems surprising that turbulences do not occur more commonly at a time of such radical change over in the circulation Many transient murmurs may originate from temporary valvular incompetence or stenosis rather than from a septal defect or patent ductus arteriosus One practical point to be remembered is that the murmurs of semilunar stenosis are the ones noted earliest in infancy If in a patient with congenital heart disease a murmur was noted at birth the chances are good that pulmonary or aortic stenosis is present

The probability that a systolic murmur heard at birth indicates congenital heart disease is 1 in 7 if the character and intensity are disregarded No figures are available in regard to the ultimate fate of newborns with "significant murmurs my impression is that no more than half of the infants prove eventually to have congenital heart disease I have personally observed the disappearance of grade 3 to 4 harsh left sternal border systolic murmurs within the first one or two months of life Whereas systolic murmurs are rarely encountered at birth by one year of age the incidence rises to 7 per cent [38] Figures quoted by various authors pertaining to normal children and adolescents vary between 30 and 50 per cent The vast majority of these correspond to the group previously labeled insignificant"

The typical commonly significant systolic murmurs are the following (1) The diamond shaped stenotic" murmur of semilunar stenosis consists of high intensity and medium frequency vibrations beginning with the first sound and lasting through most of systole Aortic stenosis murmur is best registered at the second right interspace and the apex of the diamond is usually at the first third of systole (Fig 106 *A*) The murmur of valvular pulmonary stenosis in contrast, is best heard at the second or third left interspace and the apex of the diamond is in mid systole (Fig 106 *B*) (2) The decrescendo murmur of mitral regurgitation

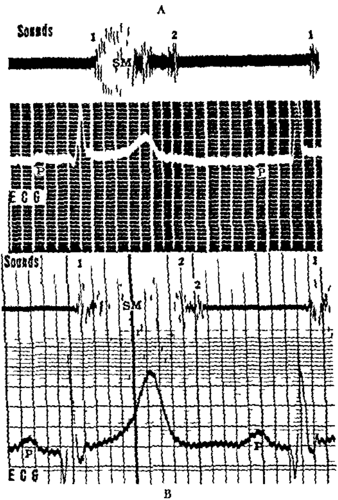


Figure 106 A Diamond shaped systolic murmur of aortic stenosis Note the early apex of the diamond a considerable distance before the second sound B Diamond shaped systolic murmur in patient with pulmonary stenosis Note the relatively late apex of the diamond close to the second sound

(Fig 107) consists of medium intensity and medium frequency vibrations and is best registered at the apex (3) The plateau shaped systolic murmur of ventricular septal defect (Fig 108) with medium intensity high frequency vibrations is best demonstrated at the third or fourth left interspace (4) The low frequency low amplitude irregular systolic murmur of the foramen secundum type of atrial septal defect is best heard at the second or third left interspace (Fig 109)

On the whole apical systolic murmurs transmitting to the left lung base are suggestive of rheumatic mitral disease and are less common in children with congenital heart disease On the other hand patients who

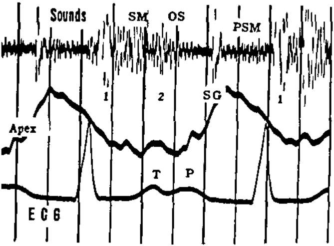


Figure 107 Decrescendo systolic murmur of mitral regurgitation Upper tracing logarithmic phonocardiogram Middle tracing apex or linear cardiogram Lower tracing electrocardiogram (From Ongley P A Sprague H B and Rappaport M B New England J Med Vol 253)

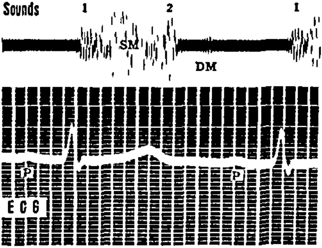


Figure 108 Plateau shaped systolic murmur of ventricular septal defect

have congenital heart disease are more likely to have left or right sternal border systolic murmurs

The typical diastolic murmurs may be described in the following terms (1) The early diastolic murmur of semilunar incompetence consists of medium amplitude high frequency vibrations They start immediately with the second sound and after an occasional short crescendo phase the major portion of the murmur is clearly decrescendo in configuration This type of murmur is best registered with a Bowles diaphragm at the left sternal border (Fig 110) (2) The early and mid diastolic low frequency low amplitude vibrations characteristic of

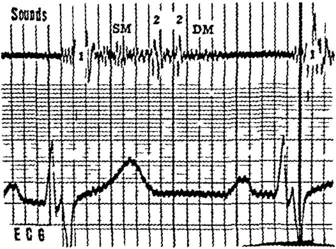


Figure 109 Irregular low intensity systolic murmur of patient with atrial septal defect Note also widely split second sound and low frequency early diastolic murmur

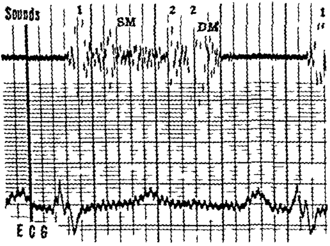


Figure 110 Early diastolic murmur of semilunar incompetence in patient with pulmonary vascular obstruction syndrome Note also widely split second sound and systolic murmur

congenital heart defects with a large left to-right shunt (atrial septal defect ventricular septal defect or patent ductus arteriosus) may register best at the left sternal border the apex or halfway between the two with the open bell chest piece (Fig 111) (3) The mid diastolic presystolic murmur of mitral stenosis (Fig 112) consists of rather low frequency low intensity vibrations giving it a rumbling character The murmur usually starts with the third heart sound gains in frequency as well as intensity toward the end of diastole and culminates in the loud

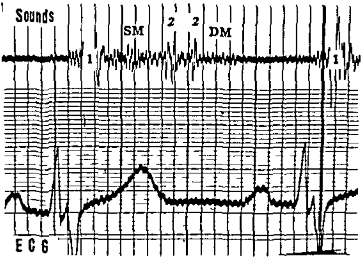


Figure 111 Early low frequency diastolic murmur in patient with atrial septal defect

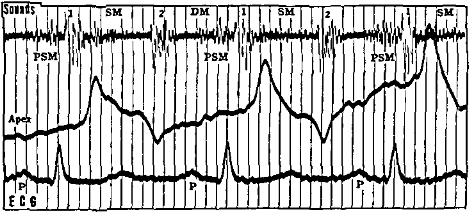


Figure 112 Typical presystolic crescendic murmur in a patient with rheumatic heart disease and mitral valve involvement Tracing recorded at cardiac apex Note (1) prominent first heart sound *1* (2) moderately split second heart sound *2* (3) decrescendic medium frequency moderate intensity systolic murmur *SM* (4) mid diastolic murmur *DM* (5) presystolic crescendic murmur *PSM*

late first heart sound This gives the picture of the typical presystolic crescendo of mitral stenosis which is a consequence of the forceful left atrial contraction and therefore is never present in patients with atrial fibrillation This murmur can best be heard with an open bell chest piece at the apex often only in the left lateral position and sometimes only after exercise (4) The continuous machinery murmur" of patent ductus arteriosus (Fig 113) is best registered at the second left interspace It consists of high frequency varying intensity vibrations and starts in a crescendic fashion with the first sound reaches its zenith at the time of the second sound and then assumes a decrescendo configuration It does not necessarily have to cover the entire cardiac cycle but in typical cases it extends a considerable distance after the second sound The

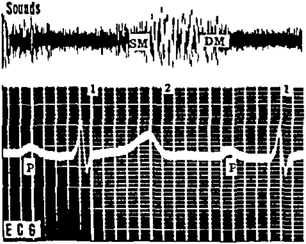


Figure 113 Phonocardiogram of typical patent ductus murmur

murmur of patent ductus arteriosus is best registered with a Bowles diaphragm

CHAPTER 5

Cardiac Catheterization

The introduction of cardiac catheterization a little more than ten years ago as a research tool and diagnostic test revolutionized the *entire field of cardiology* Concepts that had been familiar only to physiologists became common knowledge to medical students as well as to practitioners of medicine The use of the intracardiac catheter made it possible to determine accurately the cardiac output in man by applying the Fick principle It enabled physiologists to compare the output of the systemic with the pulmonary ventricle thus shunts from left to right and right to left could be quantitated with reasonable accuracy By determining the oxygen content of the blood in various *segments of the right side of the circulation the site of left to right* shunts could definitely be localized in most instances By accurate analysis of the pressure tracings stenotic lesions of the right side of the heart were discovered and pulmonary hypertension was established as a rather common disease By paying particular attention to the pressure tracing in the pulmonary tree important conclusions were drawn in relation to diseases affecting the left side of the heart

These are only a few of the great contributions of this technique to the diagnosis and treatment of heart disease Almost more important is the fact that a type of physiologic thinking was introduced into everyday cardiology which brought this branch of medicine nearer to the basic science laboratory The publications of Cournand Bing and Dexter contain the basic information necessary for the understanding of this complicated technique[3, 86, 90]

WHO SHOULD PERFORM CARDIAC CATHETERIZATION AND WHY?

Of course all this was not an unmixed blessing The question of who should be responsible for the operation of a cardiac laboratory became a real problem The doctor many years and many textbooks away from the physiology courses at medical school had to relearn" all that he might never even have understood previously The practitioner—especially the pediatrician who in the past had felt intellectually rather superior if he could distinguish the P wave from the T wave in the electrocardiogram—was suddenly given the task of analyzing the difference between the left atrial and the pulmonary capillary pressure tracings The medical man studiously avoiding the use of the scalpel since

he had finished that rotating internship he had never wanted to take to begin with had to learn how to cut down on veins and poke around the tricuspid valve with a long rubber tube The surgeon whose distaste for the stethoscope fully matched the horror of the scalpel experienced by his colleagues in internal medicine had to rediscover mathematical equations and the use of a slide rule and even to relearn Ohm s law One solution to the problem was to turn the whole procedure over to a doctor trained primarily in the basic sciences or even to a Doctor of Philosophy who could make good use of his technical skill acquired in the experimental laboratory and his intellectual superiority carefully cultivated by the various meetings of learned societies This sounded fine to everyone but the patient who still wanted to see a doctor *primarily trained in the care of people with interest in him as a person* and not only as a composite of pressures and flows

Ten years after the introduction of the cardiac catheterization technique this problem has not been solved with finality Different centers throughout the country have utilized people with different types of training for the operation of the cardiac catheterization laboratory It seems clear to me at least that the best results have been obtained in those centers where various types of personnel are available to utilize fully the benefits derived from the procedure A doctor well trained in physiology has to be in immediate and complete charge of the laboratory he should be responsible for the analysis of the data and their interpretation and significance in relation to the particular study A cardiologist should be in charge of the patient He should decide subject to the approval of the laboratory head who should undergo the procedure He should integrate the physiologic picture and the clinical observations and should interpret the whole thing to the patient and the family Finally taking the entire situation into careful consideration he should decide about the therapeutic implications subject of course to the approval of the surgeon In addition the active participation of members of the Anesthesia Service and the Radiology Department is indispensable It cannot be emphasized strongly enough that all these people should be in close cooperation and consultation with each other for best results

INDICATIONS AND RISKS

There is no unanimity of opinion regarding the indications for cardiac catheterization A great deal depends on the experience and temperament of the physician making the decision

Before embarking on the indications it must be realized that *this is a major procedure which may terminate through arrhythmia or embolism in the death of the patient* This may not be a statistically significant fact nevertheless it should be made crystal clear to every patient or parent If death occurs as a consequence of a therapeutic procedure the relatives usually accept it much more readily than if the catastrophe is

encountered as the outcome of a diagnostic procedure I do not believe that we can quote statistically valid mortality figures since much depends not only on the skill of the operating team but also on the material with which they are working If the physician is conservative and reserves catheterization for sick people only his mortality figures will be higher than those of his colleague who catheterizes everybody with an apical systolic murmur But—and this point can scarcely be overemphasized—some of the latter group the ones with no significant disease at all can be seriously harmed and may even die during the procedure The risk quoted to parents in our department is that about 299 out of 300 of these procedures go perfectly well but once in about 300 times trouble arises Higher risks than these are quoted when patients with pulmonary vascular obstruction or Ebstein's disease are considered for study

Always remembering these dangers we recommend cardiac catheterization for the following groups of patients (1) patients who on the basis of clinical evidence do not have operable heart disease but whose condition is clearly severe and in whom an operable situation cannot be excluded with reasonable certainty (2) patients with a clinical picture of operable heart disease in whom the severity of the lesion represents part of the operative indications (atrial septal defect pulmonary stenosis mitral stenosis) and (3) patients with a clinical picture of operable heart disease in whom a complicating factor possibly contraindicating operation is suspected (patent ductus arteriosus or coarctation of the aorta with right ventricular hypertrophy)

We do not usually recommend catheterization for patients who clearly have operable heart disease with definite indications for operation (severe mitral stenosis patent ductus arteriosus coarctation of the aorta or tetralogy of Fallot) We are also reluctant to use the procedure in patients without significant heart disease

The preferred age for cardiac catheterization is five years or older However successful right heart catheterization has been accomplished in many babies less than six months of age when clear indications existed

The difficulties encountered consist principally in the development of arrhythmias For obvious reasons atrial premature beats or even paroxysmal atrial tachycardia is not nearly as worrisome as are ventricular premature beats or ventricular tachycardia

In addition to the arrhythmias we have also seen transient hemiplegias presumably due to air embolism develop after catheterization Another complication we witnessed was the looping of the catheter in the right atrium with the formation of a knot cardiotomy had to be performed in order to withdraw the catheter from this patient Two more complications to be mentioned are (1) blood loss due to excessive withdrawal which may have to be corrected by postoperative blood trans

chemical laboratory completely equipped for Van Slyke determinations oxygen analysis and so forth should be available In addition to this basic equipment many laboratories have oximeters bicycle pedals for exercise tests and various other instruments furnishing supplementary information.

INFORMATION OBTAINED

The information obtained at catheterization relates in essence to two points the oxygen content and pressure level in various segments of the circulation Figure 116 lists the oxygen consumption the oxygen content

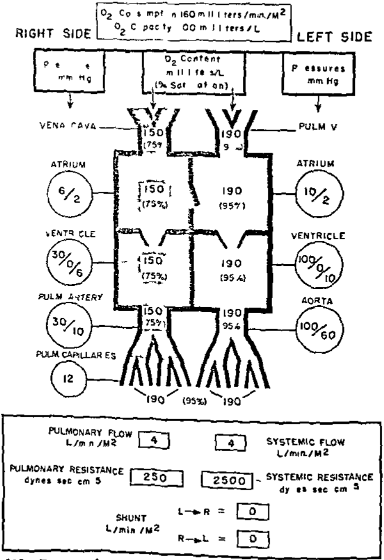


Figure 116 Diagram of normal circulatory dynamics with pressures oxygen contents and per cent of saturations

and pressure measurements with the pertinent calculations in healthy people

Calculation of Cardiac Output

Correlating the results of the oxygen analysis with the oxygen consumption the cardiac output can be calculated The basis of these calculations is the Fick principle If we can determine how much oxygen is used by the organism within a given period for example one minute and if we can also ascertain at cardiac catheterization how many cubic centimeters of oxygen are extracted from each liter of arterial blood during the same period then it is easy to calculate how many liters of blood have to go around every minute to give up the number of cubic centimeters of oxygen determined by the oxygen consumption test.

$$\text{Cardiac output L./min} = \frac{O_2 \text{ consumption cc./min}}{\text{A V difference cc./L.}}$$

The cardiac output is expressed in liters of blood per minute The oxygen consumption is expressed in cubic centimeters per minute and can be determined by making the patient breathe directly into a Douglass bag or on the basis of a table computed according to weight and height *The arteriovenous oxygen difference is determined by analyzing for* oxygen blood samples from a systemic artery (brachial or femoral) and from a part of the venous system where presumably complete mixing of the venous blood had taken place (in patients without cardiac shunts this sample is obtained from the pulmonary artery) Two examples illustrate the whole calculation clearly

$$(1)\ \text{Cardiac output} = \frac{O_2}{\text{A V difference}}\ \text{(see Fig 116)}$$

If O_2 consumption = 160 cc./min

O_2 content of arterial blood = 190 cc./L

O_2 content of mixed venous blood (P A) = 150 cc./L

$$\text{then C O} = \frac{160 \text{ cc./min}}{190 \text{ cc./L} - 150 \text{ cc./L}} = \frac{160 \text{ cc./min}}{40 \text{ cc./L}} = 4 \text{ L./min}$$

Obviously comparison of the cardiac outputs of patients of various sizes can be made only by relating the actual value to that for a standard sized person By convention a patient having a body surface area of 1 square meter is the standard means of comparison The cardiac output expressed in L./min./M² is what we call the cardiac index (C I) The cardiac index is determined by relating the cardiac output to the patient's calculated surface area As is seen in the next example if a patient's cardiac output is 3 liters and his body surface area is 1 5 square meters the cardiac index is 2 L./min./M²

$$(2)\ \text{C O} = \frac{150}{190-140} = 3 \text{ L./min}$$

Body surface area = 1 5 M^2

C I = 2 L./min./M^2

The calculation of the cardiac output by the method described is accurate to about 10 per cent and is one of the most significant results of cardiac catheterization It finds its most important application in the study of congestive failure

Determination of Cardiac Shunts

The next pertinent application of the data obtained by cardiac catheterization is in the determination of cardiac shunts Since no great change in the oxygen content of the blood in the various chambers of the right side of the heart occurs under normal circumstances (Fig 116) significant increase in the oxygen content in one chamber as compared with another means an addition of oxygenated blood from the systemic circuit Since the mixing of the venous blood is least complete in the right atrium the changes in this chamber must be relatively large to be significant Conversely since the venous blood in the right ventricle is rather completely mixed even small changes in the pulmonary artery are significant In other words if the right atrial blood sample contains 1 volume per cent more oxygen than the sample from the superior vena cava this does not necessarily indicate the presence of a cardiac shunt at the atrial level In contrast if the pulmonary artery sample is 1 volume per cent higher than the right ventricular sample that is good presumptive evidence for the presence of a patent ductus arteriosus In our laboratory the significant increase in individual right heart chambers is defined as at least 1 5 volumes per cent increase in the right atrium at least a 1 volume per cent increase in the right ventricle and at least a 0 5 volume per cent change in the pulmonary artery If these increases are found the presence of arteriovenous shunts at the respective levels can be assumed with reasonable certainty A significant increase at the right atrial level indicates an atrial defect or anomalous pulmonary vein at the ventricular level it means a ventricular septal defect and at the pulmonary artery level a patent ductus arteriosus or an aortopulmonary fenestration Individual examples of catheter findings at various shunt levels will be presented in the section on Congenital Heart Disease

A second and even more definite indication of an intracardiac communication is the passage of the catheter from the left to the right side of the heart through a defect Entrance into a left sided chamber may be recognized topographically under the fluoroscopic screen chemically by finding blood samples with oxygen content identical to that in the systemic artery and manometrically if pressure tracings characteristic of the left side of the circulation are obtained It may easily be imagined however that under certain circumstances all three of these criteria will be necessary to ascertain whether the catheter is in the left or the right side of the heart

The final contribution of the catheter to the shunt problem is the information it reveals about the magnitude of the transfer from one side to another The size of the shunt as well as its site may be calculated

with reasonable accuracy if the blood flows in a left to right direction—from systemic to pulmonary circuit In contrast if the principal flow of blood is from right to left often only the size of the transfer can be estimated without any accurate information as to the level where it occurs

The magnitude of the left to right *shunt can easily be calculated by* recognizing the fact that the cardiac output of the two sides is not identical in these cases because although the oxygen consumption is the same the arteriovenous difference is not By applying the Fick principle to the systemic and pulmonary circuits separately and by subtracting the output of one ventricle from that of the other the size of the shunt can be determined with reasonable accuracy

Two examples readily illustrate the situation

(1) O_2 consumption = 150 cc min Hence C O (systemic) = $\frac{150}{190-140}$ = 3 L /min

BA O content = 190 cc /L

C O (pulmonary) = $\frac{150}{190-160}$ = 5 L

RV O_2 content = 140 cc /L
PA O_2 content = 160 cc /L

Shunt from left to right = 2 L /min at the pulmonary arterial level

O capacity = 200 cc /L
PV O content = 190 cc /L (assumed)

(2) O_2 consumption = 150 cc /min Hence C O (systemic = $\frac{150}{170-120}$ = 3 L

BA O_2 content = 170 cc /L

C O (pulmonary) = $\frac{150}{190-120}$ = 2 1 L /min

PA O content = 120 cc /L
O capacity = 200 cc /L

Shunt from right to left = 0 9 L /min level not determined

PV O content = 190 cc /L (assumed)

It may be seen in example 1 that the systemic output is smaller than the pulmonary hence the shunt is from left to right and its magnitude is two thirds that of the systemic output It is also clear from this example that the shunt occurs at the pulmonary arterial level hence it indicates the presence of a patent ductus arteriosus

In contrast in example 2 the systemic output is about 50 per cent more than the pulmonary output implying a right to left shunt the site of which cannot be determined from these particular data It may be seen that in order to calculate the systemic flow in a patient with right to left shunt the assumption has to be made that the pulmonary vein is 95 per cent saturated

In cases of bidirectional shunts by means of more complicated equations both left to right and right to left shunts can be estimated simultaneously

Determination of Pressures

The third and perhaps most accurate group of data obtained by means of catheterization of the right side of the heart relates to the

The calculation of the cardiac output by the method described is accurate to about 10 per cent and is one of the most significant results of cardiac catheterization It finds its most important application in the study of congestive failure

DETERMINATION OF CARDIAC SHUNTS

The next pertinent application of the data obtained by cardiac catheterization is in the determination of cardiac shunts Since no great change in the oxygen content of the blood in the various chambers of the right side of the heart occurs under normal circumstances (Fig 116) significant increase in the oxygen content in one chamber as compared with another means an addition of oxygenated blood from the systemic circuit Since the mixing of the venous blood is least complete in the right atrium the changes in this chamber must be relatively large to be significant Conversely since the venous blood in the right ventricle is rather completely mixed even small changes in the pulmonary artery are significant In other words if the right atrial blood sample contains 1 volume per cent more oxygen than the sample from the superior vena cava this does not necessarily indicate the presence of a cardiac shunt at the atrial level In contrast if the pulmonary artery sample is 1 volume per cent higher than the right ventricular sample that is good presumptive evidence for the presence of a patent ductus arteriosus In our laboratory the significant increase in individual right heart chambers is defined as at least 1 5 volumes per cent increase in the right atrium at least a 1 volume per cent increase in the right ventricle and at least a 0 5 volume per cent change in the pulmonary artery If these increases are found the presence of arteriovenous shunts at the respective levels can be assumed with reasonable certainty A significant increase at the right atrial level indicates an atrial defect or anomalous pulmonary vein at the ventricular level it means a ventricular septal defect and at the pulmonary artery level a patent ductus arteriosus or an aortopulmonary fenestration Individual examples of catheter findings at various shunt levels will be presented in the section on Congenital Heart Disease

A second and even more definite indication of an intracardiac communication is the passage of the catheter from the left to the right side of the heart through a defect Entrance into a left sided chamber may be recognized topographically under the fluoroscopic screen chemically by finding blood samples with oxygen content identical to that in the systemic artery and manometrically if pressure tracings characteristic of the left side of the circulation are obtained It may easily be imagined however that under certain circumstances all three of these criteria will be necessary to ascertain whether the catheter is in the left or the right side of the heart

The final contribution of the catheter to the shunt problem is the information it reveals about the magnitude of the transfer from one side to another The size of the shunt as well as its site may be calculated

BRACHIAL ARTERIAL TRACING

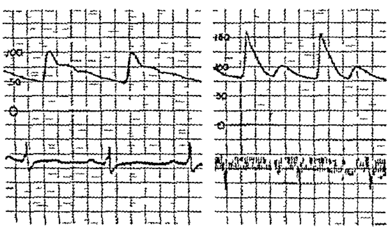


Figure 119 Comparison of brachial arterial tracing of normal person with that of patient with aortic regurgitation Note the rapid upstroke and rapid downfall of the aortic regurgitation tracing and the absence of the dicrotic arch

elevation of right atrial mean pressure is seen in right sided failure The a wave in the right atrial tracing is exaggerated (Fig 117) in severe pulmonary stenosis with intact ventricular septum pulmonary vascular obstruction and tricuspid atresia The tall V waves are characteristic of tricuspid regurgitation Right ventricular diastolic pressure is elevated in right sided failure Right ventricular systolic pressure is high in pulmonary valvular or vascular obstruction with a drop in pressure from right ventricle to pulmonary artery in the former (pressure gradient) (Fig 118)

An elevation of the pressure in the right atrium and an identity of this with right ventricular and pulmonary arterial diastolic pressure are found in constrictive pericarditis or myocardial disease Elevated left atrial and pulmonary capillary pressure is found in left sided heart failure and mitral stenosis Low diastolic pressure in the systemic artery is seen in aortic regurgitation and patent ductus arteriosus (Fig 119)

Other Calculations

Orifice sizes (mitral valve pulmonary valve) may be calculated by considering the blood flow the pressure gradient and the duration of that part of the cycle when blood flows through the particular valve (Gorlin s formula)[14]

By modifying Poisseuille s equation the resistance in any part of the circuit can be calculated from the pressure gradient and the blood flow The results are expressed in dynes seconds and centimeters to the minus fifth power and have principally a comparative value

CHAPTER 6

Angiocardiography

Angiocardiography is the visualization of the heart chambers by serial roentgenograms during the injection of an iodine-containing contrast material into the circulation By means of serial exposures taken at intervals of a fraction of a second one may trace the course of the opaque substance through successive heart chambers and blood vessels Information may thus be obtained about the size shape and location of individual heart chambers and the great vessels In addition if the venous system is injected (angiography) the presence of a right to left shunt may be visualized or if the dye is injected into the aorta from the carotid arteries (aortography) a left to right shunt through a patent ductus arteriosus may be revealed

TECHNIQUE AND EQUIPMENT

The technique consists in introducing the contrast material at maximal speed into the venous or arterial system and obtaining a maximal number of roentgenograms of the heart while the dye traverses the various chambers and thoracic vessels A great many strides in technique have been made since the introduction of the method by Castellanos in 1937 [9]

In venous angiography the dye is introduced by means of a cannula through a cut down" into an antecubital vein (or a femoral in infants) In so called selective angiography an intracardiac catheter is introduced into a right heart chamber or pulmonary artery and the dye is injected directly into the catheter If the dye is injected through a cannula directly into the left carotid artery an aortogram is obtained

The contrast material used in our institution is 70 per cent Diodrast or Urokon for venous injection and a 35 to 50 per cent concentration of the same preparations for purposes of aortography The amount of contrast material required varies with the size of the child and the size of the heart on the whole 1 cc per kilogram of body weight is sufficient for most situations On the basis of this dosage scale 8 to 10 ml are used for infants less than one year of age 10 to 15 ml are usually satisfactory for those between one and two years and 15 to 25 ml are administered for older children These figures are designed for patients with moderate cardiac enlargement larger hearts may need more dye

Rapid manual injection of the contrast material introducing the total

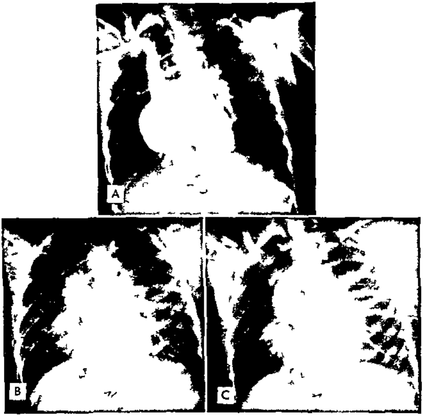


Figure 1.0 Course of the dye injected in the right arm through the chambers of a normal heart *A* The superior vena cava right atrium right ventricle and the large pulmonary artery filled *B* Incipient filling of the left atrium and left ventricle through the pulmonary veins *C* No opacification of the right side of the heart, but good visualization of the left ventricle and aortic arch

amount in less than two seconds is usually satisfactory for conventional angiography or aortography To obtain maximal benefits from the most rapid casette changers utilized in selective angiography high pressure mechanical dye injectors are used they are capable of injecting a large amount of dye in less than one second

The first angiocardiograms were obtained with a manual casette changer taking a total of four to six films in one plane every one or two seconds Today the faster machines are capable of obtaining roentgenograms in two planes at right angles at a speed of 10 per second or more By using an image amplifier and a high powered movie camera, cinefluorographic angiocardiograms that is moving pictures of the course of the dye have been obtained recently in this country and in Sweden [91]

A light general anesthetic usually Vinethene combined with pure oxygen is used for the duration of the injection to spare the patient discomfort and to avoid movement during exposure time If the arterial



Figure 121 Selective injection of dye into right ventricular outflow tract Upper picture posteroanterior view Lower picture lateral view Note in the upper picture the marked dilatation of the main pulmonary artery and in the lower picture the out line of the pulmonary valve

system is injected directly or if venous angiography is carried out on a patient with a large right to left shunt the carotids should be firmly compressed bilaterally by the anesthetist flooding the brain with Dio drast may have disastrous results

Within the framework of conventional venous angiography an outline of the heart chambers especially on the right side is obtained (Fig 120) and right to left shunts can be demonstrated By "selective" injection of the right ventricle detailed outline of the right ventricular outflow tract and the pulmonary valve may be obtained (Fig 121) Retrograde aortography may demonstrate the site of a coarctation of the aorta (Fig 122) or the presence of a patent ductus arteriosus (Fig 123)

INDICATIONS

Patients with cyanotic congenital heart disease in whom the site of the right to left shunt cannot be clearly established by clinical methods are preferred subjects for venous angiography Thus the technique is of



Figure 122 Retrograde injection into the left carotid artery outlining a coarctation of the aorta

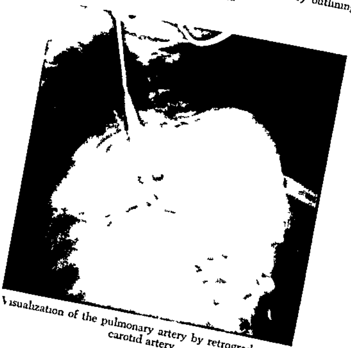


Figure 123 Visualization of the pulmonary artery by retrograde injection of the left carotid artery

considerable assistance in the differential diagnosis of the tetralogy of Fallot as against a single ventricle with pulmonary stenosis pulmonary stenosis with a foramen ovale or tricuspid atresia By demonstrating an overriding aorta communicating with both ventricles the tetralogy of Fallot may also be differentiated by this method from a complete transposition of the great arteries in which the aorta takes off exclusively from the right ventricle Truncus arteriosus communis gives a typical picture by angiocardiography in the posteroanterior view

Venous angiography usually furnishes only equivocal evidence in the diagnosis of left to right shunts

Selective angiography is of considerable help in clearly outlining the type and the site of pulmonary stenosis—specifically whether the block is at the infundibular or the valvular level or possibly at both sites *By injecting the dye directly into the pulmonary artery* through an intracardiac catheter in patients suspected of having pulmonary venous anomalies the point of entry of these anomalous veins may successfully be demonstrated the surgical importance of this can scarcely be overrated

Retrograde aortography finds its principal use in outlining the site and the length of coarctation of the aorta and in demonstrating the presence of a patent ductus arteriosus or aortopulmonary fenestrations

The enthusiasm with which angiocardiography is used varies just as much from center to center as do the indications for venous catheterization At The Children's Medical Center in Boston at the present time angiocardiography is used in only about 5 per cent of the patients with congenital heart disease admitted to the Hospital for diagnostic purposes We use it almost exclusively in four types of situations (1) atypical patent ductus arteriosus (2) cyanotic congenital heart disease (3) pulmonary venous anomalies and (4) occasionally in coarctation of the aorta

DANGERS

The dangers of angiocardiography much like those of cardiac catheterization depend a great deal on the indications for its use Published reports based on series involving several thousand cases quote the mortality rate as possibly 1 in 300 to 400 in cyanotic children and 1 in 1500 in noncyanotic patients The principal dangers are respiratory arrest, hypotension and circulatory collapse anaphylaxis and convulsions

In order to minimize the dangers we recommend that any one examination be limited to one injection multiple injections carry higher risks To avoid anaphylactic reactions angiocardiography should not be performed on a person with a history of allergy A preliminary skin test is advocated as a matter of legal technicality as much as for screening

PART TWO

ACQUIRED HEART DISEASE

CHAPTER 7

Acute Rheumatic Fever and Rheumatic Heart Disease

ACUTE RHEUMATIC FEVER

INCIDENCE AND ETIOLOGY

Rheumatic fever is one of the two most important causes of heart disease in children It is difficult to determine its incidence accurately since there is no unanimity as to diagnostic criteria Furthermore the disease is not a reportable one Nevertheless using patients with rheumatic heart disease as a principal source of information it must be concluded that rheumatic fever is common In our climate about 25 per cent of the adults with heart disease can clearly be classified as rheumatic[354] In England in 1927 it was estimated that 10 to 15 per cent of children less than twelve years old were affected by rheumatic fever or rheumatic heart disease[463] School surveys in the United States within the past ten years place the incidence of rheumatic heart disease in children five to nineteen years of age at between 2 and 6 per 1000 depending on geographic location[166] Finally according to Metropolitan Life Insurance Company statistics in 1944 rheumatic fever and rheumatic heart disease was the principal cause of death among persons five to twenty years of age

Although proof pointing to the magnitude of the problem seems unequivocal there is equally convincing evidence that rheumatic fever has been declining in incidence in the last fifty years Glover[134] found that the crude death rate from rheumatic fever and rheumatic heart disease fell from 67 per million of population in 1901 to 22 per million in 1937 in England There has been an equally steady decline in rheumatic mortality and morbidity in the United States during the past forty years according to the Metropolitan Life Insurance Company[166] The downward trend has increased significantly within the past fifteen years The same tendency may be discovered in statistics available from the United States armed forces in World War II the incidence of rheumatic fever was only one tenth of that in World War I Whether this change has

cipally in verrucose endocarditis localized on the atrial surfaces of the mitral valve and the ventricular surfaces of the aortic valve The tiny kernels consisting of debris and fibrin are found predominantly at the line of closure of the valve and on the chordae tendineae The valves are swollen edematous and distorted The parietal endocardium is rarely involved The myocardium is the site of perivascular round cell infiltration and specific Aschoff bodies these are usually seen in the subendocardial myocardium especially in the left atrium and the ventricles The pericardium if involved in the process characteristically presents the picture of serofibrinous pericarditis varying from the "bread and butter" type to the predominantly exudative form

The development of these acute or subacute changes furnishes the anatomic basis of rheumatic heart disease The verrucose lesions at first easily removable from the valves become more and more adherent The valves themselves lose their elasticity or become scarified and the leaflets adhere to each other The papillary muscles and chordae tendineae thicken and become plastered with exudate If there are recurrences in the rheumatic process fresh vegetations and fresh exudates are produced irregularly on the already affected surfaces As a result of the thickening and adhesion of the leaflets stenosis of the mitral valve occurs within a matter of years (two to eight years according to Cary Coombs[84]) However individual cases of mitral stenosis have been known to develop in less than one year's time The shortening of the chordae and the papillary muscles prohibits the valves from closing completely hence they become incompetent This process in contrast to the stenosis may develop (at least in a functional sense) within a few weeks

Aschoff bodies the principal feature of myocardial involvement in the rheumatic state are short lived Within a few weeks or months they are transformed into scar tissue hence an Aschoff body in the myocardium probably indicates that the rheumatic process is relatively active If scar tissue develops around the conduction system permanent conduction disturbance may result If the myocardial involvement is severe and persistent, serious and even fatal cardiac dilatation may ensue without major valvular deformity

Rheumatic pericarditis although of severe prognostic importance in the acute stage seldom leads to chronic pericardial disease Constrictive pericarditis on a rheumatic basis seldom if ever occurs If the heart can recover from the endocardial and principally the myocardial involvement the pericardium usually heals without any significant residuum

Other organs involved in the rheumatic process should be mentioned briefly In the acute stage the lungs may be the site of pneumonitis of the hemorrhagic type Rheumatic encephalitis with round cell infiltration around the basal ganglia has been described The rheumatic joint is characterized by redness of the synovia, serous fluid accumulation in the joint spaces and fibrin deposits over the cartilage Aschoff bodies may

be found in the periarticular tissue and the synovia Rheumatic subcutaneous nodules attached to tendons or periosteum may be found over joints scalp and vertebrae Histologically they represent granulation tissue similar to Aschoff bodies Nodules usually last several weeks or months [39] They can be produced experimentally by injecting blood subcutaneously into rheumatic patients

CLINICAL PICTURE

Since as has been pointed out before rheumatic fever is considered to be a specific host reaction to invasion by the group A hemolytic streptococci it can easily be seen that the clinical picture depending on the host will be protean indeed Depending on age heredity climate and economic factors a number of rheumatic syndromes may present themselves

Antecedent Infection In about 50 per cent of children having rheumatic fever the parents describe a cold a sore throat or even scarlet fever ten to fourteen days prior to the onset of illness This was treated either with an inadequate dose of an antimicrobial agent or only transiently The severity of this respiratory infection was average and its duration usually no more than two to four days

The Acute Attack GENERAL SIGNS OF INFLAMMATION These usually begin ten to fourteen days after the antecedent infection and consist of (1) fever characteristically this is not high it ranges between 101 and 103 F and is of the relapsing type High spiking temperatures such as are seen commonly in rheumatoid arthritis are rare (2) Polymorphonuclear leukocytosis of moderate degree (3) elevated erythrocyte sedimentation rate (4) lassitude irritability and pallor out of proportion to the mild anemia and moderate weight loss

SPECIFIC MANIFESTATIONS These may best be discussed under the headings of individual syndromes recognizing the fact that commonly not one but several of the syndromes of varying intensity make up the clinical profile

Arthritis Rheumatic fever involves several joints simultaneously or in succession Monoarthritis almost never is a manifestation of rheumatic fever The joints involved are usually the large ones knees ankles wrists and elbows Heat redness swelling severe pain and tenderness are the rule If as is the custom on many orthopedic services the joint is tapped clear serous sterile fluid of high protein and cell content is obtained From this description of rheumatic arthritis it may be seen how sharply different it is from the vague muscle and tendon pains called "growing pains the relation of which to rheumatic fever is questionable Differentiation of the two conditions is possible by simply asking the mother what she is doing for the pains of the youngster If she says that the child likes to have his limbs rubbed or massaged one can be almost certain one is not dealing with rheumatic arthritis This condi

tion in its true form makes the joints so tender that even the weight of a blanket may be intolerable

Carditis Definite evidence of carditis almost requires the diagnosis of rheumatic fever in children Although accurate figures on the occur rence of carditis in unselected cases of rheumatic fever are not available the incidence is usually considered to be at least 50 per cent and it is found in every fatal case

For the evaluation of carditis a knowledge of the condition of the patient s heart prior to the onset of the present illness is of great im portance This is particularly pertinent in trying to determine whether or not the presence of a murmur indicates acute carditis If the murmur was clearly not present before the attack of rheumatic fever the likeli hood that it signifies carditis is strong Apical systolic murmurs can be considered significant only if they last at least through the first half of systole and if the intensity is at least grade 3 Apical diastolic murmurs always significant are commonly found in acute carditis at a time when no organic mitral stenosis is present[39] The origin of this murmur is not clear but it is thought to be due to relative mitral stenosis caused by dilatation of the left ventricle Aortic diastolic murmurs are also noted *early in the history of carditis in contrast to the diastolic murmurs in* the mitral area these are more likely to become permanent All these murmurs may develop into permanent auscultatory features or may be entirely transient and the heart after recovering from acute carditis may sound entirely normal

The heart sounds in acute carditis often lose their luster and become distant dull and muffled Taran's[9] emphasized the development of embryocardia an equalization of the duration of systole and diastole resulting in a tic tac like rhythm Diastolic gallop is commonly present

Cardiomegaly of appreciable degree can usually be demonstrated on physical examination Serial radiograms give objective evidence and are useful as guides to prognosis At fluoroscopy the heart beat is usually feeble Enlargement of the left atrium demonstrated in the right anterior oblique view is emphasized by Wilson[38] as a prime criterion of carditis

Pericarditis can usually be detected by the presence of a friction rub Sometimes with the development of a large effusion the friction rub disappears at this stage however the massive cardiomegaly the feeble heart *sounds poor cardiac pulsation and evidences of tamponade usu* ally reveal the diagnosis On the other hand rheumatic pericarditis is practically never seen without a significant murmur Although peri carditis is of serious prognostic significance it usually does not require paracentesis except occasionally for diagnostic purposes

Evidences of congestive failure that is distended neck veins puffy face hepatomegaly pulmonary rales low urinary output and even pitting edema can all be regarded as evidences of carditis Almost axiomatically any child with rheumatic heart disease who is in failure

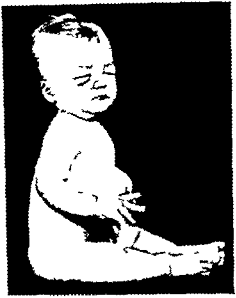


Figure 124 Photograph of little girl with congestive failure due to rheumatic carditis Note the similarity to the features of patients with nephrosis

must have active carditis This is in marked contrast to older people in whom congestive failure may be the outcome of the mechanical stress placed on the heart by rheumatic valvular involvement *Right sided failure particularly involving facial edema is characteristic of congestive failure in rheumatic children* Pure left sided failure on the other hand is relatively rare The accumulation of fluid in some of these youngsters may be so marked that they have been known to be admitted to hospital wards as "nephrotics" (Fig 124)

In discussing the characteristic features of active carditis in addition to the specific signs of congestive failure—endocardial, myocardial or pericardial involvement—the less specific but nevertheless characteristic general signs and symptoms must also be mentioned These children are anxious frightened and perspiring they show slight peripheral cyanosis which improves in oxygen The pulse is usually rapid and thready although bradycardia has been found in 50 per cent of a series of adults with rheumatic fever [167] Dyspnea orthopnea with flaring of the alae nasi is prominent

The electrocardiogram is a sensitive index of active carditis [65] The changes may be transient, however and unless frequent even daily electrocardiograms are taken may be missed completely Prolongation of the P R interval (first-degree block) and the presence of the so-called Wenckebach phenomenon (second degree block) (Fig 125) are the most commonly mentioned abnormalities However *the presence of a prolonged P R interval per se does not mean active rheumatic fever* [65]

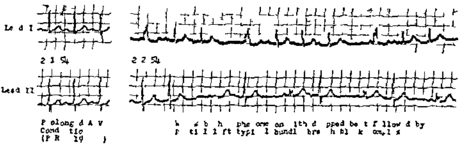


Figure 125 Electrocardiographic changes in acute rheumatic fever Note in the tracing of February 1 the prolongation of the P R interval in the tracing of February 2 the Wenckebach phenomenon with left bundle branch block or aberrant interventricular conduction

Many cases of impaired atrioventricular conduction reflect only past rheumatic activity in others it may be congenital in origin In this respect a transient prolongation of the atrioventricular conduction time is more suggestive of an active process than is a permanent one We have even seen second degree block persist through the years without any evidence of active rheumatic fever Taran[340] emphasized the importance of measuring the corrected Q T interval in cases of suspected rheumatic fever he regarded a QTc of 0 42 second or more as definite evidence of active carditis He also stated that with obvious carditis a QTc of less than 0 42 second probably indicates pericardial involvement These observations have been confirmed by Wood[363] but others have failed to find a high degree of correlation between active carditis and QTc[88, 118] Nodal rhythm S T and T wave changes atrial fibrillation and bundle branch block are less common evidences of active carditis Sinus tachycardia is the rule as mentioned with occasional instances of normal rate or even sinus bradycardia

Chorea In terms of immediate prognosis chorea is the most benign of the rheumatic syndromes whereas all patients who die with acute rheumatic fever have carditis practically none has chorea There has also been noted an antagonism between the presence of chorea and the other acute rheumatic syndromes the simultaneous occurrence of polyarthritis and chorea is practially unknown In spite of this there can be little doubt that chorea is one of the rheumatic manifestations since according to various authors from 50 to 75 per cent of patients with chorea exhibit other rheumatic manifestations at some time prior to or following its appearance According to the well controlled series of Ash[13] about a third of these patients have definite rheumatic heart disease ten years after the onset of the chorea

St. Vitus dance is different from the other rheumatic syndromes not only because it is a solitary event within the rheumatic cycle but also because of the absence of general signs of inflammation (fever leukocytosis) and of acute phase reactants (sedimentation rate C reactive protein)

The occurrence of an antecedent infection is more than usually difficult to determine because of the vague onset of the disease Emotional disturbances are usually the first manifestations The child most often a girl between the ages of eight and twelve years becomes difficult to live with She tends to be extremely moody and irritable At school she cannot concentrate at home she refuses to do her chores She fights incessantly with her siblings Parents and teachers alike resent her attitude and pressures are put on This only aggravates the situation the changes in mood become truly explosive

A few weeks after the gradual onset of emotional instability involuntary movements are first noted They are quick and brusque and involve all parts of the body Voluntary movements become difficult to execute and bring on choreiform twitchings The handwriting deteriorates and becomes almost illegible Glasses and china are easily broken when the child helps with the dishes She cannot unbutton her clothes or lace her shoes

Signs of ataxia appear The speech becomes slurred and gait hesitating her movements lose all their grace and become jerky and angular When sticking out her tongue at the time of the physical examination she jerks it back suddenly and snaps her jaws The tongue itself may show twitchings and fibrillary motion

Evidences of weakness occur Her grasp is hesitating and unsustained She cannot play her favorite games and becomes tired easily

All these signs of weakness and locomotor ataxia may be limited to one side of the body only (hemichorea)

Skin Manifestations The rashes seen in patients with rheumatic fever may include nonspecific lesions such as urticaria petechiae and erythema nodosum On the other hand erythema marginatum is for all practical purposes a specific and main manifestation of severe rheumatic fever It consists of a circinate macular and erythematous rash appearing on the trunk or the extremities but not on the face usually symmetrically distributed and of varying duration The typical pattern consists of a wavy thin red line surrounding an area of intact skin about 1 to 2 cm in size (Fig 5 p 13) There is no itching or discomfort associated with it

Rheumatic nodules are definitely a major and specific manifestation of rheumatic fever Similar to erythema marginatum they occur almost exclusively in the severest types of rheumatic fever with carditis They are never transient if untreated their duration is from several weeks to months They occur symmetrically singly or in clusters loosely attached to joint capsules tendon sheaths or the periosteum They are freely movable under the skin Most commonly they are seen over the joints scalp and vertebrae Initially they may best be discovered by looking for them in an indirect light later they can be felt without any difficulty

They are not painful or tender their size varies from that of a small pea to a small almond (Fig 4 p 13)

Epistaxis Severe nosebleeds commonly occur in acute rheumatic fever although within the past decade their incidence at The House of The Good Samaritan in Boston has definitely been decreasing Repeated nosebleeds in a child with rheumatic heart disease but without any of the major manifestations of the disease are not especially rare

Abdominal Pain Often the presenting symptom of a patient with acute rheumatic fever is abdominal pain If this is accompanied by general signs of inflammation a rigid abdomen and right lower quadrant localization the differential diagnosis from acute appendicitis may be difficult indeed If an exploratory laparotomy is done the peritoneum will be found to be moderately injected with swollen mesenteric glands and a slight increase in the peritoneal fluid Acute appendicitis is the *admitting diagnosis in a number of patients with acute rheumatic* fever referred to our hospital

Pneumonia Rheumatic pneumonia is rare and indicates severe rheumatic fever Furthermore it may be impossible to tell in any given case how many of the signs and symptoms of pneumonitis are caused by left sided congestive failure pulmonary hemorrhage secondary infection or true rheumatic granulation tissue

Differential Diagnosis

Jones in a historic paper in 1944 [160] suggested definite diagnostic criteria for rheumatic fever Although later he stated that publication of a set of definite criteria might introduce an element of unnecessary rigidity into the diagnosis of rheumatic fever still for didactic purposes this represents an extremely useful classification He divided the clinical syndromes into two groups major and minor manifestations The major manifestations were carditis chorea subcutaneous nodules and previous attacks of rheumatic fever The minor manifestations were fever abdominal pain precordial pain rashes epistaxis pulmonary changes anemia and high sedimentation rate later an elevated antistreptolysin O titer was added to the list Jones felt that the diagnosis of rheumatic fever could be made with certainty if any two major or one major and two minor manifestations of rheumatic fever were present The least specific of these combinations is the presence of arthralgia with fever and elevated sedimentation rate

Recently a committe of the American Heart Association modified these criteria in the following manner * major criteria (1) carditis (2) polyarthritis (3) chorea (4) subcutaneous nodules (5) erythema marginatum Minor criteria (1) fever (2) arthralgia (3) prolonged

Jones Criteria (Modified) for Guidance in the Diagnosis of Rheumatic Fever Report by the *Committee on Standards and Criteria for Programs of Care Circulation* *13* 617 1956



Figure 126 Typical joint involvement of patient with rheumatoid arthritis

P R interval in the electrocardiogram (4) increased erythrocyte sedimentation rate presence of C reactive protein or leukocytosis (5) evidence of preceding beta hemolytic streptococcal infection (6) previous history of rheumatic fever or the presence of inactive rheumatic heart disease

Although in many instances the diagnosis of rheumatic fever can be made at the time of the first visit in a number of cases observation extending to weeks or months is necessary before a definite opinion can be expressed On the other hand since the advent of hormone therapy a number of investigators especially Massell [38] and his associates at The House of The Good Samaritan have stressed the necessity of early diagnosis They believe that steroids in adequate doses administered early in the disease—before the advent of carditis—may prevent the development of valvular deformities Since among the five major manifestations arthralgia is the only one that is likely to precede carditis by an appreciable interval (chorea is usually not followed by carditis within the same cycle and rheumatic nodules without carditis are extremely rare) a great deal of emphasis is placed on the early identification of arthritis as rheumatic Thus the presence of polyarthritis with signs of inflammation and an elevated antistreptolysin titer should be regarded as prima facie evidence of rheumatic fever It is difficult to argue against the validity of this concept in terms of diagnosis However the therapeutic implications to be drawn from it represent in my mind a much more controversial point

Probably all children in whom the diagnosis of rheumatic fever has been made on the basis of the criteria mentioned will have the disease The only exceptions are patients having other collagen diseases which are really cousins of rheumatic fever Rheumatoid arthritis or Still s disease

should be mentioned in detail Children with rheumatoid arthritis may have either no fever at all or a spiking septic type of temperature Even if the fever is high the patients are usually not sick in proportion to their febrile response The joints involved are more commonly the smaller ones (metacarpal or metatarsal) or the unusual ones (sternoclavicular or mandibular) The inflammatory process is not as painful as that which we are accustomed to see in rheumatic fever the joints are tender but not excruciatingly so and are only moderately red The involvement is more chronic with fluid accumulation lasting for weeks and months in the same joint The affected fingers assume a spindly appearance (Fig 126) Cardiac involvement is rare if it occurs it is more likely to attack the pericardium and the myocardium leaving the endocardium intact although aortic regurgitation may occur in adults Thus murmurs are rather rare Pulmonary arterial hypertension caused by pulmonary vascular disease has been seen in patients with rheumatoid arthritis Splenomegaly is fairly common Fleeting rashes of the erythema multiforme type are seen momentarily erythema marginatum is rare Laboratory tests reveal mild anemia marked leukocytosis and elevated sedimentation rate The antistreptolysin titer may or may not be elevated

As can be seen from this brief description the border between rheumatoid arthritis and rheumatic fever is not sharp It is my personal belief that they may represent different host reactions to the same insult everyone who has dealt with this problem to any extent has had the experience of seeing a patient with a clear cut diagnosis of one of these conditions turn into an equally characteristic picture of the other Nevertheless from the practical and principally prognostic viewpoint the two entities should be kept separate It may be said that rheumatoid arthritis is chronic joint disease without verrucose endocarditis

Disseminated lupus erythematosus and periarteritis nodosa should also be mentioned as members of the collagen disease group in which widespread visceral and vascular lesions dominate arthritis and carditis playing only minor roles

A list of the referring diagnoses of patients who later turned out to have definite rheumatic fever may give some idea of the other problems in differential diagnosis No detailed description of the distinguishing points will be given simply thinking about the entities will serve the purpose appendicitis poliomyelitis toxic synovitis osteomyelitis nephritis leukemia tuberculosis and septicemia

Course and Prognosis

Rheumatic fever is a cyclic disease Both the general signs of inflammation and the specific syndromes may wax and wane through weeks months and perhaps years Individual cycles are usually three to six weeks apart Between these although the disease seems quiescent the sedimentation rate stays high no significant weight gain occurs and

1000 Patients						
Initial	347 No heart disease			653 Heart disease		
20 years follow up	4/7 without heart disease	3/7 with heart disease	10 died	1/6 without heart disease	2/6 with heart disease	3/6 died

Figure 127 Prognosis of acute rheumatic fever (From E F Bland and T D Jones Rheumatic Fever and Rheumatic Heart Disease A Twenty Year Report on 1000 Patients Followed since Childhood Circulation Vol 4)

anemia persists The patient does not seem to be quite himself and the sleeping pulse rate may be high It takes several months before one can be entirely certain that the disease has subsided There is no single criterion to indicate when the acute attack has ended Most centers taking care of large numbers of rheumatic patients require that a number of criteria be satisfied before they announce the process as being arrested The principal requirements are as follows (1) no evidence of major manifestations of rheumatic fever (2) no signs of inflammation (3) satisfactory weight gain (4) normal erythrocyte count and hemoglobin and (5) general sense of well being good disposition and good appetite

With the termination of the acute attack whatever endpoint one may use the question of recurrences and ultimate prognosis should be considered The figures published by Bland and Jones from The House of The Good Samaritan reflect the history of this disease as seen in Boston between 1920 and 1950[37, 181] The conclusions based on this twenty year follow up of 1000 patients should not however be taken as an indication of what the outlook is now or what it was fifty years ago The severity of individual attacks has shown a definite decline within the past half century and the number of recurrences has been kept considerably lower within the past decade The following data on recurrences and long term prognosis are based on the figures of Bland and Jones

The number of recurrences in their study depended a great deal on the time which had elapsed after the first attack During the first five years there was a 20 per cent probability for recurrence during the second five years the figure dropped to 10 per cent during the third five years to 5 per cent and finally between fifteen and twenty years after the original attack only 1 4 per cent had recurrences

The long term prognosis in the 1000 cases reviewed by Bland and Jones is summarized in Figure 127 At the end of the first attack of rheumatic fever 65 per cent of 1000 patients had definite rheumatic heart disease Of these children with definite heart disease a third died

within the first ten years and another sixth during the next ten yea To put it in another way half of those who emerged from their fi attack of rheumatic fever with heart disease were dead twenty yea later Most of these deaths occurred within five years after the init attack Ash stated[14] in fact that more children die within the first ye of their disease than in any subsequent year

In contrast to the poor prognosis of the group who acquire hea disease at the time of their first attack stands the favorable outlook of tl 35 *per cent who escape manifest cardiac damage initially* Of this grou of 347 patients none died as a consequence of rheumatic fever or rhe matic heart disease *in the next twenty years*

Reviewing these mortality statistics according to specific rheumat syndromes it becomes obvious that gross cardiomegaly and congesti failure have the worst prognostic implications 70 to 80 per cent of tl patients with these conditions died within the first ten years In contras only 12 per cent of the children whose initial syndrome was chore died within the same period

The cause of death was acute rheumatic fever and congestive failu in 80 per cent and subacute bacterial endocarditis in 10 per cent of tl cases

Of the total 1000 patients about 650 were still alive twenty yea after the initial attack and three fourths of these were able to lead relatively normal existence

Somewhat less than half of the survivors (316 patients) had n residual heart disease at all Two thirds of these were patients wh escaped from their first attack without heart disease however a third c them (108 patients) were thought at the end of their first attack to hav definite evidence of rheumatic heart disease on the basis of apical sy tolic or mid diastolic murmurs and even an occasional instance of fair aortic regurgitation In Ash s series[13] a carefully executed follow up stud indicated that about 10 per cent of the survivors of acute carditis abou 50 per cent of the survivors of polyarthritis and 66 per cent of th survivors of chorea had no residual heart disease ten years after the onse of rheumatic fever

Of the 650 patients in Bland and Jones series who survived twent years 383 had definite rheumatic heart disease The majority of thes had acquired heart disease during their initial attack However abou 150 of them mostly patients with chorea had not had heart diseas initially in these mitral stenosis developed insidiously without over evidence of recurrence through the years

Treatment

Until the last four or five years no specific treatment for acut rheumatic fever was available *There is still no unanimity in regard to the effectiveness of various drugs on acute rheumatic fever* but an increasing

number of clinicians believe that the course of the disease may be significantly influenced by a number of therapeutic agents The use of hormones salicylates anticongestive measures and other therapeutic approaches will be discussed

Hormones The demonstration by Hench and his associates[164] in 1949 of the beneficial effects of ACTH and cortisone therapy on patients with arthritis and other hypersensitivity states raised the hope that a cure for rheumatic fever might be at hand

In order to make a scientific evaluation of the effects of the hormones on acute rheumatic fever a combined study group was organized in the United States Canada and Great Britain This group included most of the large centers for study of rheumatic diseases in the respective countries and employed the talents of first rate investigators and a host of statisticians A rigid schedule for case selection dosage and length of treatment was set up Within a relatively short time 500 cases were collected a third of these patients were treated with aspirin another third with ACTH and a final third with cortisone The three groups were selected in such a fashion that statistically significant comparisons could be made

A detailed report of this study has been published recently[56] indicating that there was no statistically significant difference in the results of the three plans of treatment Aspirin seemed even slightly more beneficial than the hormones and the pendulum swung away from hormone treatment

While all this was going on members of the study group made their own observations without the aid of IBM machines Other investigators throughout the United States Canada and England drew their own conclusions without statisticians as doctors have done since time immemorial The almost unanimous conclusion was that, from the clinical viewpoint the hormones were immensely useful indeed Though nobody could say with certainty that residual heart disease was less likely to develop in the hormone treated patients almost everybody felt that the sickest patients the ones with pancarditis and congestive failure were much more likely to recover with the use of the new agents than they would have been under the old treatment plan There also seemed to be unquestionable and startling effects on fever sedimentation rate polyarthritis pericarditis subcutaneous nodules and electrocardiographic changes No significant effects on chorea or erythema marginatum were noted The survey of the independent observations to date on the treatment of acute rheumatic fever with hormones has been admirably summarized by Massell[239]

The opinion of most observers today is that hormones are a useful tool in the treatment of acute rheumatic fever Obviously they are not the whole answer to the problem but the clinical impression prevails that the sickest children are much more likely to recover with hormones

than they did before the advent of the steroid era The conviction is growing also that the disappointing results of the combined study may have been due to late administration of probably inadequate amounts for too short periods

On the basis of our own experience at The House of The Good Samaritan and the data presented in the literature I believe that the sickest children with rheumatic pancarditis and congestive failure the group which showed an 80 per cent mortality in Bland and Jones series [37, 181] have an excellent chance to survive with the aid of hormones today I do not expect that the hormones will eliminate existing heart disease I do believe however that there is good circumstantial evidence that hormones given early enough long enough and in adequate doses may suppress the inflammatory process in the heart to a sufficient degree to prevent the development of valvular damage Massell [38] and his associates demonstrated in a convincing fashion that rheumatic nodules not principally different from rheumatic granulation tissue elsewhere disappear rather rapidly under the influence of cortisone or ACTH Thus it does not seem too farfetched to assume that this may happen in the heart as well if irreversible scarring has not yet taken place

For these reasons I would consider the presence of severe pancarditis with congestive failure a definite indication for the use of hormones In this situation they may be lifesaving and I would not withhold them A less definite but probably valid indication for the use of these agents is the presence of acute rheumatic fever with incipient carditis (murmurs electrocardiographic changes cardiomegaly tachycardia) but without congestive failure In these patients if hormone therapy is started early one may expect that evidences of carditis will be quickly suppressed the development of valvular damage will be prevented or minimized and the length of hospitalization will be shortened It is my personal belief that one is fully justified in using steroid treatment for these types of patients but I am aware that this belief is based more on faith and impressions than on factual data

For a third group of patients those with acute rheumatic fever without clinical evidence of carditis I do not feel justified at this time in recommending hormone treatment According to Bland and Jones [37, 181] two thirds of their patients without initial carditis did not exhibit heart disease twenty years later The patients in Bland and Jones group by virtue of the fact that they were referred to The House of The Good Samaritan must have represented a collection of more seriously ill children than the ones the pediatrician is likely to see in his private practice If heart disease did not develop in two thirds of these patients who were perhaps relatively sicker than the average and who did not later benefit from chemoprophylaxis one is scarcely justified at this point in recommending *hormone treatment for the average patient with acute* rheumatic fever who does not have cardiac involvement It seems to me

Table 9 Dosage Schedule for Steroid Treatment of Acute Rheumatic Fever

Period of Treatment in Days	Total Daily Dose in Mg			
	Meticorten		Oral Cortisone	
	Over 40 lbs	Under 40 lbs	Over 40 lbs	Under 40 lbs
21	60	40	300	200
7	50	35	250	175
7	45	30	225	150
7	40	25	200	125
7	35	20	175	100
7	30	15	150	75
7	25	10	125	50
5	20	5	100	25
5	15	2 5	75	12 5
5	10		50	
6	5		25	

that more careful studies on the subject must be made before all rheumatic fever patients irrespective of carditis can be treated with hormones In patients with chronic recurrent rheumatic fever the hormones like anything else available at this time are probably of no use

Treatment should start early there is considerable evidence that treatment instituted shortly after the initial attack has a much better chance to succeed than treatment administered later Dosages should be high and should last for at least twelve weeks A tentative schedule for cortisone and Meticorten used presently at The House of The Good Samaritan is included in Table 9

A useful adjunct in the hormone treatment of acute rheumatic fever is careful control of the fluid intake to no more than 1000 cc per day The diet should contain no more than 50 to 100 mg of sodium chloride daily Potassium chloride in doses of 1 5 to 2 gm should be added to the diet Vigorous anticongestive measures especially mercurials and digitalis should be used if congestive failure is present Antibiotics probably penicillin or one of the broad spectrum drugs should be administered to guard against intercurrent infection and to eradicate any streptococci present in the nasopharynx

There are definite drawbacks and dangers to hormone treatment Cushing s syndrome (Fig 128) with considerable weight gain will develop in all patients treated according to the schedule proposed This will retrogress in almost all cases but it is an unpleasant sight while it lasts Severe acne may develop in the ACTH treated group In some patients significant arterial hypertension develops to the extent that treatment cannot be completed Glycosuria may appear psychoses although not common in children do occur The fluid retention inherent in steroid



Figure 128 Patient with fully developed Cushing's syndrome

treatment may prove to be disastrous to patients with congestive failure unless the beneficial effects of the hormones on the myocardium compensate for this

One further drawback to hormone treatment is the development of the so called rebound phenomenon This consists in the appearance of a flurry of signs and symptoms of activity when hormones are discontinued There is no good explanation for this phenomenon The only way it can be *differentiated from true return of rheumatic activity is by observing* the patient for a few days perhaps for a week At the end of this period the manifestations of "rebound" should spontaneously disappear

Salicylates For many years salicylates have been administered to achieve symptomatic relief in acute rheumatic fever About ten years ago Coburn[83] *attempted to prove that intravenous administration of* salicylates in doses raising the salicylate level to at least 30 mg per 100 ml of blood influences the course of the disease itself This thought although alien to the thinking of English and American physicians has been widely accepted in Europe for a considerable time There is still no conclusive proof of the therapeutic effect of salicylates in rheumatic fever there is no question of their value in symptomatic management However Coburn's data and the results of the combined study [86] suggest that these agents are therapeutically effective The consensus today is that when salicylates are administered they too much like the hormones should be given in large enough doses for a long enough time

The dosage used today at The House of The Good Samaritan is 1 grain (0 06 gm) per pound of body weight per day in four to six divided

doses The total dose should not exceed 10 gm per day This dosage should be continued for at least two weeks when gradual tapering off may be introduced Salicylate treatment should be continued however for at least six weeks or until all signs of activity have disappeared When salicylates are administered in large doses vitamin C and vitamin K should be given simultaneously to prevent hemorrhage If evidences of salicylate intoxication appear the dose may have to be diminished or the drug entirely discontinued Among the salicylate preparations aspirin seems to be as effective as any The enteric coated tablets of sodium salicylate are palatable but their satisfactory absorption cannot always be assumed

Salicylates are the drugs of choice today for patients having acute rheumatic fever without carditis except in cases of chorea in which even symptomatically they are not effective Whether salicylates should be used in addition to hormones in patients with severe carditis is debatable

Anticongestive Measures The general principles of management of congestive failure will be discussed in a separate chapter (see p 242) *Rheumatic fever patients in congestive failure should receive anti congestive drugs in addition to steroids in essentially the same fashion as do other patients with congestive failure*

A great deal of controversy surrounds the use of digitalis in rheumatic carditis Much of the argument centers around the point that a certain number of patients with acute rheumatic carditis die when digitalis is administered to them [303, 304] Often it is forgotten that some of these patients are extremely sick and might die irrespective of the treatment I believe that cautious electrocardiographically controlled administration of digitalis is an extremely useful tool in the treatment of congestive failure associated with rheumatic carditis Of course it must be remembered that these children may be extremely sensitive to the glycoside and doses smaller than those usually given will be optimal in their treatment It also should be pointed out that potassium depletion may develop in patients treated with corticoids especially if mercury is administered simultaneously and potassium intake is low it is well known that digitoxicity is more prone to develop in patients with low serum and intracellular potassium levels [3] Consequently if digitalis is given to a patient treated with steroids an adequate potassium intake must be assured

Eradication of the Streptococcus The streptococcus carrier state can easily be treated with penicillin Oral administration of one million units of penicillin per day in four divided doses for ten days or the intramuscular injection of 600 000 units of depot penicillin every other day for four doses accomplishes the purpose in practically all instances After the eradication of the streptococcus penicillin prophylaxis should be started immediately Sulfonamides are apparently relatively ineffec

tive in the treatment of streptococcal infections, although they may be used effectively for prophylactic purposes

Bed Rest For the duration of active carditis bed rest must be absolute the child should be fed and washed and must use a bed pan After subsidence of evidences of carditis the patient still must stay in bed but may be allowed to wash and feed himself Bathroom privileges are first given when all clinical evidences of activity disappear Gradually after all the laboratory and clinical evidences of activity have subsided the patient may be allowed out of bed for several hours a day The minimum amount of time to be spent in bed for any patient with acute rheumatic fever is probably two months No patient with carditis should get out of bed for at least four months should return to school for six months or should resume full activity for a year Prolonged care for this length of time usually is best accomplished in a rheumatic fever convalescent home

Diet The use of salt restriction in the diet of patients with congestive failure and those under hormone treatment has already been mentioned During the feverish stage children do not want to eat and they are better off with a liquid carbohydrate diet When the fever has subsided *especially if they are under steroid treatment* the appetite will improve They should be given a diet high in protein carbohydrates and vitamin C The appetites of some of the hormone treated patients are perfectly astounding

PROPHYLAXIS

The use of chemotherapeutic agents for the prevention of rheumatic fever is predicated on the acceptance of two premises one is the etiologic role of group A beta hemolytic streptococci the other is the significance of the number of recurrences in the prognosis of rheumatic fever If as most people are willing to accept today recurrent rheumatic fever damages the heart and if recurrences are dependent on the presence of hemolytic streptococci then it seems logical to try to prevent streptococcal infections thus avoiding rheumatic recurrences and precluding further cardiac damage

I believe there is little argument against this type of theoretic reasoning However considerable controversy surrounds two points about the practical application of these principles One concerns the duration of chemoprophylaxis and the other the use of prophylactic measures in patients with throat infections who have not previously had rheumatic fever

If the figures of Bland and Jones[37, 161] regarding the relationship of rheumatic recurrences to the time elapsed since the original infection are valid there seems to be no good reason to administer chemoprophylaxis longer than ten years after the first attack Nevertheless even if no more than 8 per cent of the patients have recurrences after ten years is it not

worth preventing those? All that one may say is that every physician must make an individual decision in regard to each patient There is a psychological point in the life of many patients at which getting rid of pills" may be worth risking something in terms of rheumatic fever Others do not mind taking medication ad infinitum Fortunately for the pediatrician this decision must be made by the internist in most instances As far as pediatricians are concerned anybody who has had acute rheumatic fever should be protected against streptococcal infection throughout childhood probably for at least ten years

The second controversial point concerns the treatment of sore throats in patients who have not had a previous attack of rheumatic fever Every pediatrician sees or talks on the telephone to the parents of hundreds of children with sore throats with or without fever I do not believe that for practical purposes it would be feasible to treat all of these for ten days with penicillin The patient s patience the druggist s bill and the psychological havoc are all limiting factors A little common sense is extremely useful in this type of situation Factors favoring the use of chemotherapy with ordinary sore throats are (1) family history of rheumatic fever (2) evidence that the throat infection is streptococcal in origin (positive throat culture presence of exudate fever leukocytosis cervical lymphadenitis) and (3) crowded living conditions that is boarding schools army camps and so forth If none of these situations exists I do not recommend that sore throats should be treated with antibacterial agents

One further point should be disposed of briefly It has been stated and I could scarcely disagree more strongly that all febrile illnesses at their onset should be treated with penicillin as a sort of diagnostic procedure If the fever subsides within twenty four hours it was probably due to the hemolytic streptococcus thus according to this line of reasoning the penicillin should be continued for ten days If the fever does not disappear I presume one ought to call a doctor to find out what is the matter with the patient

Any pediatrician knows that fevers of unknown origin and of no particular consequence make up a vast majority of his daily practice he also knows that bed rest fluids and mild antipyretics will keep the situation in hand in most instances Although it has to be granted that an occasional case of rheumatic fever may result from this symptomatic regimen it still seems to me the sensible course to follow because of the relative rarity of the disease and the untold psychological economic and even physical damage that would inevitably follow the continuous use of penicillin

The drug of choice in chemoprophylaxis at present is penicillin If a suspicion of acute streptococcal infection exists the treatment schedule previously outlined for eradication of the streptococcus carrier state should be used (see p 147) This schedule should be followed by the

prophylactic schedule The simplest way to administer prophylaxis is to give the oral tablets 100 000 units three times a day or 200 000 units twice a day Monthly injections of a depot type of penicillin (Bicillin) have also been advocated but the local reactions seem less than completely favorable

Instead of penicillin sulfonamides may be used for prophylaxis but not for treatment of streptococcal infections

Results of large scale carefully controlled investigations have shown the effectiveness of both penicillin and sulfonamide prophylaxis [193 195 40 336 34]

Recurrences of rheumatic fever in the groups using prophylaxis were about a tenth of those occurring in the control group or even less

The eradication of streptococcal infections was equally startling Well controlled studies have proved without any doubt that adequate treatment with penicillin not with sulfonamides at any stage of the streptococcal infection helps to prevent the occurrence of acute rheumatic fever [5] The earlier the treatment the better the results will be but recent studies suggest that vigorous treatment with penicillin started as late as nine days [46] after the onset of illness still will result in a significant decrease in the number of patients with acute rheumatic fever

RHEUMATIC VALVULAR DISEASE

It may be possible for the internist to separate a chapter on rheumatic fever into two divisions one dealing with the acute attack and the other with the mechanical consequences of rheumatic fever that is rheumatic valvular disease Histologic studies showing Aschoff bodies in the left atrial appendage in patients undergoing mitral commissurotomy [336] however strongly suggest that low grade active disease may be present even in adults without clinical evidences of rheumatic activity

In pediatrics it has long been known that every child having symptomatic rheumatic valvular disease should be strongly suspected of having some degree of activity This is obviously not the case in all instances but the burden of proof is on the physician who says that the rheumatic process is totally inactive in a child who is symptomatic with rheumatic heart disease Consequently it seems wiser to discuss the entire rheumatic heart disease problem under one heading In the following pages individual valvular lesions will be discussed as if they represented the only pathologic condition present although the presence of acute rheumatic fever cannot in most instances be excluded with certainty

Diseases of the Mitral Valve

Evidences of mitral heart disease are obtained through auscultation in most instances It is usually assumed that significant murmurs heard maximally at the apex of the heart are referable to the mitral valve the assumption is also made that if no murmurs are heard at the apex, the

mitral valve is intact Although as will be seen later exceptions to these rules exist still by and large they do hold true The mitral valve alone or in combination with aortic valvular disease is involved in about 90 per cent of the patients with rheumatic heart disease

According to the studies of Bland and Jones[47, 181] it may be said that evidence of mitral heart disease appearing during an attack of acute rheumatic fever means permanent involvement of this valve in about 75 per cent of the cases

Mitral Stenosis

Mitral stenosis is the most common single valvular lesion developing as a consequence of rheumatic endocarditis Women are more commonly affected than men The disease is rare in children At least two but probably more years are needed for its development A considerable number of patients with no evidence of valvular involvement in the acute attack will have mitral stenosis ten to twenty years later The stenosis may vary from only slight narrowing of the orifice (mitral valve area of 2 0 to 2 5 sq cm) to a calcified buttonhole stenosis (mitral valve area of 0 5 to 1 0 sq cm) In children calcification is extremely rare although severe stenosis may occur

PHYSIOLOGY

The principal hemodynamic difficulty in mitral stenosis is the imposition of a barrier between the left atrium and the left ventricle in diastole The basic hemodynamic principles have been well worked out by Dexter Gorlin and their associates[143, 14, 146] These physiologic studies demonstrate that in order to overcome the obstruction at the site of the mitral valve the pressure in the left atrium is raised and a gradient between left atrial mean and left ventricular end diastolic pressure is established This elevated left atrial pressure is reflected in the high "pulmonary capillary" pressure obtained at cardiac catheterization This type of compensation may be completely satisfactory for many years

With the progression of the stenosis however complications may arise from several directions First the left atrium may not be able to overcome the increasing resistance offered by the mitral valve and thus the cardiac output diminishes Second by trying to overcome the block left atrial and "pulmonary capillary" pressure is raised often to a level (more than 25 mm of mercury) at which pulmonary edema may result Finally the increased "pulmonary capillary" pressure may stimulate pulmonary vascular obstruction on an organic or reflex basis and thus pulmonary arterial hypertension and right ventricular failure may result At right heart catheterization patients with mild degrees of mitral stenosis show little that is abnormal With increasing severity of the disease the following changes may be noted "pulmonary capillary" pressure increases pulmonary arterial hypertension occurs on exercise later

even at rest cardiac output diminishes as the systemic arteriovenous dif ference increases later the cardiac output becomes fixed and finally right ventricular end diastolic pressure increases and right sided con gestive failure is present

Clinical Picture

The first heart sound at the apex is accentuated The second sound at the pulmonary area is loud only if pulmonary arterial hypertension is present An opening snap (Fig 101 p 100) corresponding to the opening of the mitral valve is commonly audible A low frequency rumbling mid diastolic murmur is audible at the apex with the progres sion of the disease this murmur extends further and further into diastole finally occupying presystole (Fig 96 p 97) where it assumes a cres cendic configuration This crescendic apical presystolic murmur often accompanied by a thrill is the pathognomonic auscultatory finding in rheumatic mitral stenosis Occasionally rarely in children a short high frequency early diastolic blow can be heard over the pulmonary area (Graham Steell murmur)

Obviously not all these phenomena are present in all cases At the onset of mitral stenosis only a loud first sound at the apex may be audible at rest This should arouse suspicion if the patient has a history sugges tive of rheumatic fever Having the patient exercise turning him in the left lateral position and listening with an open bell stethescope may bring out the diastolic presystolic murmur Later with the development of atrial fibrillation the presystolic crescendic murmur always disappears thus furnishing conclusive proof that when present it is due to the forceful atrial contraction squeezing blood through a tight mitral opening

The rest of the physical examination is less specific Patients have high color characteristically a malar flush with suggestion of peripheral cyanosis is described Probably this is due to the low cardiac output and the high oxygen extraction in the capillaries Left chest deformity usually is not marked The cardiac impulse is of right ventricular character The pulse is usually small and shows a narrow pulse pressure

The electrocardiogram in pure mitral stenosis of some severity shows right axis deviation vertical heart and right ventricular hypertrophy In adults more than forty years old or in children with active carditis atrial fibrillation frequently occurs If P waves are present they are commonly abnormal Their abnormalities consist in an increase in height and width in the standard limb leads occurrence of diphasic P waves in lead V_1 and flat topped or notched P waves in leads I and II

The radiologic appearance of mitral stenosis is characterized by evi dence of left atrial enlargement (Fig 129)

Most of the so called complications of mitral stenosis are seldom if ever seen in children Hence they will be mentioned only briefly (1) Atrial fibrillation presumably the effect of overdistention of the left

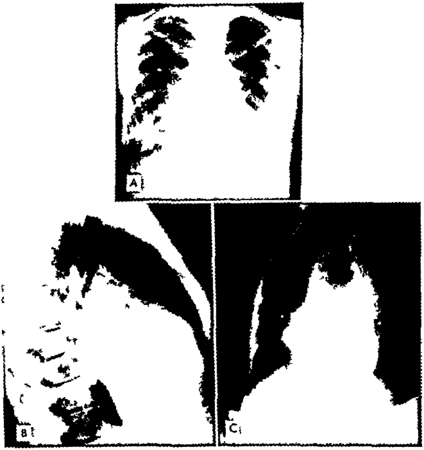


Figure 129 Radiograms of patient with mitral stenosis

atrium occurs frequently in patients more than forty years of age If it is observed in children or adolescents it almost invariably means active carditis (2) Hemoptysis is the result of infarction or pulmonary capillary obstruction Occasionally hemoptysis occurs in very sick patients with acute carditis and "rheumatic pneumonia" (3) Bacterial endocarditis always should be thought of in conjunction with rheumatic heart disease but actually its occurrence is rare in pure mitral stenosis especially in children

Differential Diagnosis

Mitral stenosis is usually easy to recognize in children a recent history of rheumatic fever can be obtained more frequently than in adults and a good mid diastolic presystolic murmur is present almost without exception

For practical purposes the only problem in differential diagnosis is presented by other conditions which give rise to apical diastolic mur

murs Among these should be mentioned acute rheumatic carditis without organic stenosis the Austin Flint murmur of aortic regurgitation and congenital heart disease with left to right shunt especially atrial septal defect If one remembers that it takes a considerable amount of time for organic mitral stenosis to develop then the apical diastolic murmur appearing within the framework of acute carditis can be recognized as probably representing a functional" murmur The Austin Flint apical presystolic murmur associated with aortic regurgitation is almost impossible to differentiate from the murmur of true mitral stenosis except by thinking about it in patients with free aortic incompetence The apical diastolic murmur of the left to right shunt group may also be hard to differentiate from that of mitral stenosis The presence of real presystolic accentuation a diastolic thrill an opening snap and a loud first sound favor the diagnosis of mitral stenosis whereas a low frequency murmur without thrill occupying early or mid diastole in a patient with marked pulmonary vascular engorgement favors the diagnosis of congenital heart disease with a left to right shunt In the latter group if incomplete right bundle branch block is present in the electrocardiogram atrial septal defect is the likely diagnosis

One further point to be mentioned in the differential diagnosis is the separation of congenital mitral stenosis from mitral stenosis of rheumatic origin This can be done only on the basis of age If mitral stenosis occurs in an infant or very young child one must assume that it is of congenital origin

Course and Prognosis

Children with mitral stenosis unless they have acute carditis are usually asymptomatic In rare instances they may show some fatigue and exertional dyspnea According to Gorlin and his associates[145] the clinical picture of advanced mitral stenosis as seen principally in adults is the result of an interplay between the narrowing of the valve the pulmonary vascular obstruction consecutive to it and the state of the myocardium Patients in group I those with the mildest stenosis have a mitral valve area of more than 1 5 sq cm and normal pulmonary vascular resistance and show only slight if any dyspnea on exertion The cardiac output of these patients is within normal limits at rest and exercise Patients in group II are characterized by a valve area of 1 0 to 1 5 sq cm and relatively normal vascular resistance These patients have respiratory symptoms some dyspnea on exertion paroxysmal nocturnal dyspnea and hemoptysis The cardiac output is normal at rest but does not rise sufficiently on exercise Patients in group III have severe mitral stenosis (valve area of 0 5 to 1 0 sq cm) with moderately increased pulmonary vascular resistance and an impaired myocardium Although these patients show considerable dyspnea on exertion or

strenuous exercise their main trouble is weakness tiredness and easy fatigability The cardiac output is low normal at rest and practically fixed on exercise Finally group IV is made up of patients with marked mitral stenosis (valve area less than 0.5 sq cm) considerable increase in pulmonary vascular resistance poor myocardium and fixed low cardiac output These patients have dyspnea at rest gross cardiomegaly and right ventricular failure

The outlook for patients with asymptomatic mitral stenosis depends on the presence or absence of recurrences diet crowding and climatic and genetic factors On the whole the prognosis of patients with mitral stenosis is perhaps less favorable than that of those having mitral or aortic regurgitation the average age at death in a series in which chemoprophylaxis had not been used was between thirty five and forty five years [4] In fact Bland and Jones figures suggested that many patients die in acute carditis Most of the deaths in their series occurred within five or ten years after the initial attack most of those who survived the first ten years were still alive twenty years after their first attack

Treatment

The treatment of mitral stenosis consists of three principal parts The most important of these is the prevention of recurrences by means of chemoprophylaxis

The second therapeutic measure is the surgical correction of the stenosis by mitral valvuloplasty [0 1 9] Probably this is not necessary in the vast majority of children it should be advocated only if one can be certain that the symptoms are due to organic mitral obstruction rather than to myocardial factors One must also be sure that severe organic mitral stenosis and not combined valvular lesions with or without active rheumatic carditis is the principal cause of the patient s difficul ties A few successful cases of mitral surgery in youngsters between twelve and fourteen years of age have been reported in the literature [8] We had two patients less than fifteen years of age who underwent mitral valve surgery The indications for surgery are (1) definite and progressive dyspnea on exertion (2) attacks of pulmonary edema and (3) paroxysmal dyspnea with hemoptysis

The patients who benefit most from the various surgical procedures are those in whom the predominant difficulty is a mechanical block at the mitral valve [116] These patients may be recognized physiologically by the presence of a low fixed cardiac output and pulmonary hyper tension with a significant rise on exercise Conversely patients with equally severe symptoms and clinical signs of mitral stenosis in whom the main difficulty is the inefficiency of the myocardium fail to respond well to mitral valvotomy these patients also have a low and fixed

cardiac output but do not have significant pulmonary hypertension at rest or on exercise

The technical aspects of mitral commissurotomy or valvuloplasty have been described adequately in many publications [illegible] The over all mortality figures vary according to the condition of the patient the accuracy of the diagnosis and the skill of the surgeon On the whole a mortality figure of about 1 to 5 per cent may be quoted for patients without congestive failure whereas the others entail a 20 to 25 per cent mortality risk The cause of death is mostly embolic and not congestive failure Among the postoperative complications should be mentioned in addition to embolism the so called postcommissurotomy syndrome [316] This peculiar cyclic disease occurring in 15 to 20 per cent of patients after mitral commissurotomy consists of fever chest pain pneumonitis and cardiomegaly It is said to be due to reactivation of rheumatic fever this is a possible hypothesis not a statement based on unassail able facts

The patient who survives the immediate postoperative period and is kept on a strict medical regimen for a few months including prophy lactic chemotherapy and the judicious use of anticongestive measures usually improves More than 60 per cent of the survivors report consid erable symptomatic relief not always documentable by physiologic observations the explanation of the discrepancy between symptoms and physiologic data belongs in the realm of psychosomatic medicine rather than in pediatric cardiology In five year follow up studies [160] late deaths occurred in less than 5 per cent of the cases reoperation was necessary in only a few patients

The final considerations in the treatment of mitral stenosis in chil dren are the assurance of good dental hygiene adequate diet and a sensible attitude toward the disease

Mitral Regurgitation

If the presence of a "significant apical systolic murmur may be taken as definite evidence of mitral regurgitation then it must be present in almost all cases of acute rheumatic carditis As a matter of fact without the presence of an apical systolic murmur the diagnosis of rheumatic carditis rests on tenuous grounds indeed About two thirds of the patients who in their acute attack show auscultatory evidence of mitral regurgitation will have definite mitral incompetence twenty years later That mitral regurgitation as an isolated rheumatic valve lesion exists is suggested by the auscultatory findings in chronic rheu matic heart disease and is supported by the observations of surgeons whose palpating fingers in left atrial exploration occasionally encounter a regurgitant jet without evidence of mitral stenosis In contrast to these clinical and surgical experiences are the observations of pathologists who maintain that pure mitral regurgitation is almost never found at post

mortem examination Probably this means only that mitral regurgitation per se is not a fatal disease In contrast to pure mitral stenosis mitral regurgitation affects men more frequently than women

Incompetence of the mitral valve anatomically speaking results from the rigidity and shortening of the valve cusps In addition the chordae tendineae contracted through scar formation also may impede complete closure of the mitral valve during ventricular systole

Physiology

The principal difficulty in mitral regurgitation is that the left ventricle and the left atrium have to handle an increased volume of blood This is necessary since a considerable portion sometimes as much as 50 per cent of the left ventricular stroke output is dissipated into the left atrium and thus the left ventricular diastolic filling has to be increased to a point at which in spite of the loss into the left atrium the aortic flow still is in the required range At right heart catheterization usually but not invariably the pulmonary capillary" pressure tracing shows unusually tall V waves at left heart catheterization a large V wave is encountered in the left atrium An increase in right atrial and right ventricular pressure is found only if the left ventricle fails The cardiac output is moderately decreased

Clinical Picture

The signs of mitral regurgitation are much less clear cut than those of mitral stenosis As has been mentioned the principal auscultatory finding is the presence of an apical systolic murmur which probably means mitral insufficiency if (1) the murmur is grade 3 or louder (2) it transmits best along the posterior axillary line to the left base (3) it starts immediately with the first sound and lasts at least through the first half of systole and (4) it does not change with position and respiration

Other features of the physical examination are left chest deformity left ventricular type of impulse a moderately accentuated pulmonary second sound a normal first sound and normal blood pressure and pulse pressure Hoarseness due to pressure on the recurrent laryngeal nerve by the large left atrium has been noted

The electrocardiogram shows left ventricular dominance or left ventricular hypertrophy occasionally with P mitrale"

Under the fluoroscope the heart seems overactive particularly in the region of the enlarged left ventricle Left atrial and left ventricular enlargement of significant degree is usually found in the more advanced cases (Fig 130) as a matter of fact the largest left atria in rheumatic heart disease are seen with mitral regurgitation Systolic expansion of the left atrium is commonly noted The pulmonary vasculature is within normal limits unless the left ventricle fails The inaccuracy of all these clinical signs in indicating the presence of mitral regurgitation has been amply

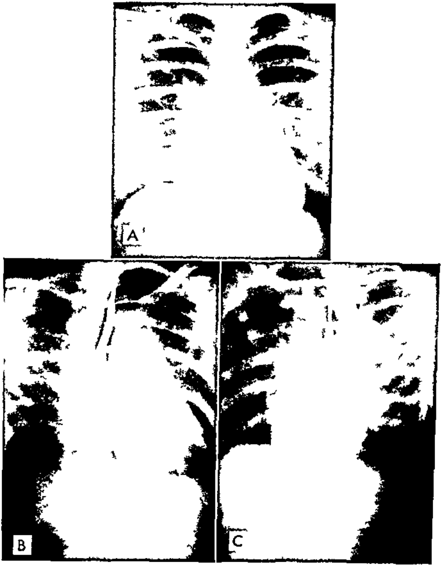


Figure 130 Posteroanterior and oblique radiograms of patient with mitral regurgitation

documented by surgeons who found significant mitral regurgitation on left atrial exploration in patients in whom none was preoperatively suspected and vice versa

DIFFERENTIAL DIAGNOSIS

All the conditions accompanied by an apical systolic murmur should be considered in the differential diagnosis of mitral regurgitation First to be considered in this regard are patients with "insignificant" apical systolic murmurs These murmurs are faint and short and

do not transmit but may change with position or respiration Using essentially the same criteria though different nomenclature Boone and Levine[4] studied two groups of patients one with "organic" murmurs the other with "functional" murmurs Ten years later the group with the "organic" murmurs contained ten times as many patients with multivalvular involvement or significant cardiomegaly as did the group with "functional" murmurs Kuttner and Markowitz,[1 4] in a similar study found that after eight years 48 per cent of the patients in the "organic" group showed significant heart disease as compared to 13 per cent in the "functional" group Although the character of the murmur is important in the differential diagnosis obviously the clinician ought to use all other tools including a good history physical examination x rays and the electrocardiogram to determine the significance of an apical systolic murmur

"Significant" apical systolic murmurs may be heard in patients with rheumatic heart disease other than mitral regurgitation It has been reported [44] that adults with aortic stenosis may show only an apical systolic murmur I have never knowingly encountered this in children

Patients with pure mitral stenosis may also have significant apical systolic murmurs which brings up the difficult question of differentiating by clinical means a predominantly stenotic type of lesion from a dominantly regurgitant one The importance of this from the surgical viewpoint can scarcely be overemphasized Fortunately for the pediatrician surgically significant mitral stenosis rarely occurs in children Nevertheless in an occasional adolescent the problem may arise It may be said unequivocally that there is no single criterion which distinguishes without fail a predominantly stenotic mitral valve from one in which regurgitation dominates A sustained left ventricular type of cardiac impulse an intense systolic murmur the presence of systolic expansion of the left atrium and an electrocardiogram indicating left ventricular hypertrophy would give the clinical impression of predominant regurgitation Although the pulmonary capillary tracing, in terms of the tall V waves is designated by some[196 317] as furnishing clear-cut proof of dominant mitral regurgitation in our hands and in the hands of others[34] this has not been a satisfactory method of differentiation Even left heart catheterization may not clearly establish the diagnosis

Congenital heart disease especially atrial septal defect of the ostium primum type and to a lesser extent patent ductus arteriosus may also be accompanied by an apical systolic murmur In both these instances this may indeed represent functional or even anatomic mitral regurgitation Finally almost any other type of heart disease acquired or congenital may be accompanied by an apical systolic murmur of "significant" nature All the ingenuity the pediatrician is able to muster

may be necessary for accurate interpretation of a grade 3 apical systolic murmur

Course and Prognosis

Most patients who have pure mitral regurgitation are relatively asymptomatic for a long time and have a good long term prognosis The symptoms—if no active carditis exists—consist mainly of fatigue Con gestive failure is rare life expectancy is satisfactory In contrast to the relatively rare occurrence of subacute bacterial endocarditis in pure mitral stenosis its presence in patients with mitral regurgitation is relatively common

About a third of the patients starting out with pure mitral regurgi tation eventually have combined stenosis and insufficiency in these symptoms develop and the prognosis becomes grave

Treatment

Treatment is essentially the same as that outlined in the section on mitral stenosis except that there is no adequate surgical solution for the problem at this time

Diseases of the Aortic Valve

Aortic valvular disease is diagnosed on the basis of murmurs heard in the aortic area that is at the second right and fourth left interspaces There is some pathologic evidence that in adults [44] aortic valve involve ment may occur without any murmur at all or with a murmur only at the apex No such observations have yet been made on children

Rheumatic aortic disease alone or in combination with mitral valvular disease is common Fifty eight per cent of the patients[37, 181] in whom rheumatic heart disease develops show evidences of aortic involvement In contrast to diseases of the mitral valve disappearance of aortic murmurs is extremely rare

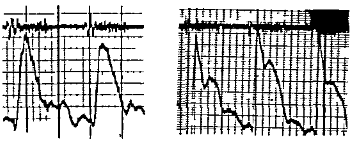
Aortic Regurgitation

Aortic regurgitation is the dominant consequence of involvement of the aortic valve in rheumatic endocarditis Men are more commonly affected than women All degrees of the disease are known from the faint early diastolic "whiff without any peripheral signs to the free aortic regurgitation represented by a loud murmur and a diastolic pressure near zero

Anatomically the valve ring is dilated the semilunar cusps are shortened by scarring and thus the valve edges fail to close the aortic orifice during ventricular diastole

Physiology

The overload of aortic regurgitation is carried almost exclusively by the left ventricle The increased volume work of this chamber is



a b

Figure 131 a Normal brachial arterial tracing and synchronous phonocardiogram on top b Aortic regurgitation tracing with a rapid upstroke and rapid downstroke As a reference tracing see also phonocardiogram on top

the consequence of the increased stroke output which is caused by the fact that a certain proportion of the blood expelled into the aorta leaks back into the left ventricle It seems clear then that the left ventricular stroke output must be large enough to insure adequate aortic flow even with appreciable loss back into the ventricle The degree of regurgitation depends on the size of the regurgitant opening and on the difference between aortic and left ventricular diastolic pressures

Significant peripheral vasodilatation is a regular concomitant of severe aortic regurgitation [3,5] Thus the drain on the aortic pressure head is from two directions part of the blood regurgitates back into the ventricle and part dissipates rapidly into the dilated peripheral vascular bed As a result of both these factors aortic diastolic pressure is low this interferes with adequate coronary filling

A low aortic diastolic pressure not only impedes coronary circulation but also interferes with the satisfactory nutrition of the tissues since it lowers the mean arterial pressure at the periphery In order to counteract the unfavorable effects of the low diastolic pressure on the mean arterial pressure the systolic pressure is appreciably raised This then results in a relatively satisfactory mean pressure with a wide pulse pressure The increased systolic pressure adds a significant amount of pressure load to the already increased volume load of the left ventricle

The high systolic and low diastolic pressure in the arterial system is reflected in the characteristic radial arterial tracing obtainable in patients with aortic regurgitation (Fig 131 see also Fig 119 p 121)

The powerful musculature of the left ventricle is able to cope with the considerable flow and pressure work involved in aortic regurgitation for an appreciable length of time Difficulties arise when the left ventricle fails or when angina develops as a result of poor diastolic filling of the coronary arteries Both these consequences occur in adult life and should affect the thinking of the pediatrician only in terms of long range prognosis rather than as events likely to occur within the first two decades of life

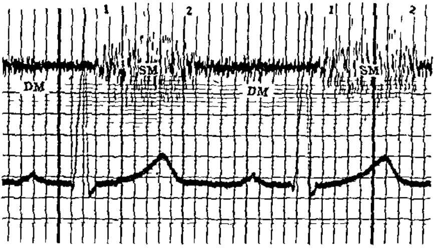


Figure 132 To-and fro murmur of aortic regurgitation Note the stenotic type of systolic murmur and the decrescendo diastolic murmur going through entire diastole

Clinical Picture

The diagnostic sign of aortic regurgitation is the early insufficiency blow which is best registered at the third to fourth left or the second right interspace This consists in high frequency vibrations immediately following the second sound in a descrescendo fashion perhaps preceded for a short period by a slight crescendic phase The stethoscope with the diaphragm type of chest piece brings out the murmur best particularly with the patient in a sitting position slightly bent forward Even though sometimes it may be almost impossible to time the murmur accurately it still should be recognized as a high frequency, metallic sounding almost "echo like murmur

The rest of the physical examination is somewhat more characteristic in these patients than in those with mitral disease Increased pulsations of the carotid arteries can be clearly seen and easily palpated the powerful and sudden thrust of the left ventricle may rock the head and chest of a patient with marked regurgitation Considerable left chest prominence may be present The cardiac impulse is forceful and of the left ventricular type Occasionally there is a diastolic thrill palpable at the third to fourth left interspace On auscultation in addition to the typical murmur described a systolic murmur of varying intensity and sometimes of stenotic character may be heard at the aortic area these two murmurs assume a to and fro character (Fig 132) with a gap between the systolic murmur and the second sound The aortic closure is usually accentuated and unless congestive failure is present it is louder than the pulmonary closure An apical presystolic murmur (Austin Flint) practically indistinguishable from the murmur of true mitral stenosis, is often observed but is not accompanied by a thrill



Figure 133 Posteroanterior and oblique radiograms of patient with aortic regurgitation

The pulses are full and the pulse pressure is wide with high systolic and low diastolic readings

The electrocardiogram may be entirely normal In severe cases of long duration left ventricular hypertrophy may be present with S T and T wave changes tall R waves in the left chest leads and P mitrale

Radiologically cardiomegaly principally left ventricular in type can be seen (Fig 133) If the left atrium is enlarged left ventricular failure or associated mitral valve disease must be suspected The aorta and the left ventricle show powerful pulsations under the fluoroscope the pulmonary artery segment is relatively quiet The ascending aorta and the aortic knob are prominent and have a collapsing type of beat

Differential Diagnosis

The differential diagnosis of aortic regurgitation encompasses two main entities One is the group of conditions with dissipation of the aortic pressure head In this category should be mentioned thyrotoxicosis severe anemia peripheral arteriovenous aneurysm patent ductus arteriosus aortopulmonary fenestration and ventricular septal defect with aortic regurgitation The first three conditions simply have to be borne in mind if the problem of differential diagnosis arises all three of them by virtue of their extracardiac findings can be recognized once they are brought to mind The three latter conditions may be much harder to differentiate from rheumatic aortic regurgitation though several points may be helpful For one thing all three of these con

genital heart diseases involve a large left to right shunt hence radiologically they are characterized by pulmonary vascular engorgement with a hilar dance and an overactive main pulmonary artery Second especially patent ductus arteriosus and aortopulmonary fenestration are accompanied by continuous systolic and diastolic murmurs where the *second sound* is enveloped into the murmur Of course a history of a murmur from the time of birth and the absence of a clear cut rheumatic episode also help to identify the left to right shunt group However in *spite of all this cardiac catheterization sometimes must be performed* in order to establish the diagnosis

The other group of patients to be considered in the differential diagnosis are those with pulmonary regurgitation (mitral stenosis pulmonary arterial hypertension and idiopathic pulmonary regurgitation) in whom diastolic murmurs of a quality similar to that of aortic regurgitation may be noted The murmur is usually less loud however it is maximal at the second to third left interspace and is not accompanied by an increase in pulse pressure

Course and Prognosis

Much like mitral regurgitation and in marked contrast to mitral stenosis aortic regurgitation may occur early within the framework of acute rheumatic fever Once an aortic diastolic murmur appears the chances of its disappearance are slim (less than 5 per cent) although there are some suggestions in the most recent literature that treatment with corticoids may change the outlook [38]

Children with free aortic regurgitation but without active carditis are usually asymptomatic The powerful left ventricle is able to maintain compensation throughout childhood and adolescence unless rheumatic myocardial disease interferes with its efficiency Severe aortic regurgitation in adults leads eventually to exertional dyspnea anginal pain pitting edema and intractable congestive failure Like those with aortic stenosis these patients "break late" but when they do their downhill course is precipitous

The severity of the lesion can probably best be judged clinically on the basis of the pulse pressure the electrocardiogram and the roentgenograms and only to a lesser degree on the basis of the intensity of the murmur The poor prognosis to be measured in terms of years must be contrasted with the good outlook and life expectancy for children with only a slight or moderate degree of aortic regurgitation The average life expectancy of patients with aortic insufficiency is quoted as twenty to thirty years after the discovery of the murmur [403]

Treatment

There is no well accepted surgical treatment for this disease Hufnagel valves [1 0] inserted in the descending aorta to abolish the effects of

peripheral vasodilatation have been used in adults At present they probably have no place in pediatric practice

Chemoprophylaxis and regulation of living conditions are just as important as they are in diseases of the mitral valve Furthermore knowing that the left ventricle is capable of maximal effort for a long time without failure but that when it fails the end is rather near I usually recommend that youngsters with aortic regurgitation abstain from competitive sports Sometimes this is hard to enforce since many of them are athletically inclined young males

Aortic Stenosis

Rheumatic aortic stenosis as an uncomplicated lesion is practically unknown in children There were only two patients in the group of 1000 studied by Bland and Jones[3] [161] in whom pure aortic stenosis developed within twenty years both these patients were more than twenty five years old

If signs and symptoms of aortic stenosis are found in a child, it must be assumed for practical purposes that it is congenital in origin The problem of aortic stenosis will be discussed in detail in the section on Congenital Heart Disease

Diseases of the Tricuspid Valve

Diseases of the tricuspid valve are rare and of no practical significance in pediatrics they almost never occur as an isolated lesion and seldom as a dominant one

Physiologically the mechanisms elaborated on in connection with mitral stenosis and mitral regurgitation operate here except that the burden falls exclusively on the right side of the heart Right atrial pressure and jugular venous pressure tracings show prominent V waves in tricuspid regurgitation and tall A waves in tricuspid stenosis In addition characteristically in cases of tricuspid stenosis there is a gradient on cardiac catheterization between right atrial mean and right ventricular end diastolic pressures

Clinically the systolic murmur of tricuspid regurgitation and the diastolic murmur of tricuspid stenosis are both heard best at the lower right sternal borders It is hard almost impossible to differentiate them from mitral murmurs transmitted from the apex Careful observation of the jugular venous pulse and the pulsations of the liver aid a great deal in the differential diagnosis Presystolic pulsations of the neck veins and the liver are characteristic of tricuspid stenosis whereas vigorous systolic pulsations and V waves of the jugular venous pulse and the liver are typical of tricuspid regurgitation Roentgenologically the huge right atrium is noted Electrocardiographically marked P pulmonale is in evidence

Combined Valvular Diseases

Involvement of more than one valve is the rule rather than the exception in rheumatic heart disease According to White[3,4] mitral diseases dominate in about three fourths of the cases and aortic valve abnormality in about one fourth Stenosis and regurgitation of the same valve usually occur together Clinically, mitral regurgitation with out stenosis is more common than mitral stenosis without regurgitation If especially in relatively acute cases one cannot be sure which mitral lesion is predominant the term "mitral involvement is useful

Aortic regurgitation without stenosis is common whereas aortic stenosis of rheumatic nature without coexisting aortic regurgitation is practically unknown in children

Bacterial Endocarditis

In contrast to the usually sterile inflammatory lesions of rheumatic endocarditis the vegetations of bacterial endocarditis contain micro organisms identifiable morphologically as well as culturally The causative organism may be cultured in vivo from the blood stream in at least 75 per cent of the patients with bacterial endocarditis blood cultures from patients with rheumatic endocarditis on the other hand are sterile

Bacterial endocarditis is divided perhaps arbitrarily into two groups subacute and acute These two disease entities can be identified by their causative organism as well as by their clinical course

SUBACUTE BACTERIAL ENDOCARDITIS

Subacute bacterial endocarditis is a slowly progressive disease in which the only or principal localization of the infection is in the heart or great vessels

INCIDENCE AND ETIOLOGY

Subacute bacterial endocarditis much like rheumatic fever has been on the decline within the past decade this trend is most likely explained by the widespread use of antibiotics for the treatment of respiratory infection and by dental prophylaxis in patients with heart disease Littman and Schaaf [18] found that whereas during the three year period from 1944 through 1946 1 12 per cent of the patients admitted to the Massachusetts General Hospital had subacute bacterial endocarditis from 1947 through 1949 only 0 59 per cent had this disease

Subacute bacterial endocarditis is a disease of young adults Men are more commonly affected than women In White s[187] series of 250 patients only 2 per cent were children less than ten years old and only 17 per cent were in their second decade Compared with the general incidence of subacute bacterial endocarditis in a general hospital its frequency at The Children s Hospital in Boston is well below 0 1 per cent

Streptococcus viridans or alpha hemolytic streptococcus is responsible for about 90 per cent of the cases of subacute bacterial endocarditis in adults Although no large series of cases in children has been described in the literature our experience at The Children s Hospital in the past ten years supports the same figure Other organisms rarely

Table 10 Pertinent Clinical Data in 12 Children with Subacute Bacterial Endocarditis

Palpable spleen	12	Anemia (other 2 severely cyanotic)	10
Fever	11	Positive blood culture	10
Petechiae	7	Increased E S R	9
Severe dental caries	9	Wbc over 20 000	6
Clubbing (3 with severe cyanosis)	6	Hematuria	4
Congestive failure	4	Prolonged P R interval	4
Hemiplegia	1	Cardiomegaly	9

The temperature most commonly is between 101 and 103° F with considerable daily fluctuation Severe dental caries is common

The presence of petechiae clinches the diagnosis although their absence does not preclude subacute bacterial endocarditis Sometimes the petechiae are elusive and look like small freckles It is worth while to mark the suspicious spots with a skin pencil and to follow them day by day to see whether they disappear or whether new ones occur It is important to note that the petechiae of subacute bacterial endocarditis are widely scattered not clustered around an area such as the neck or the antecubital fossa Localized petechial spots suggest increased capillary permeability rather than embolic phenomena Petechiae sometimes persist after bacteriologic cure of subacute bacterial endocarditis and may last for several months without ill effect

Clubbing is significant as a diagnostic aid only if the patient does not have cyanotic congenital heart disease in this situation however it is practically pathognomonic if liver disease and lung abscess can also be excluded

Congestive failure especially right sided may be the first manifestation of subacute bacterial endocarditis in a patient with known heart disease On the other hand congestive failure may begin after bacteriologic cure of the endocarditis In the latter instance it is usually blamed on the myocardial damage caused by the infection Transient hemiplegia with or without convulsions in a patient with known heart disease should raise the suspicion of subacute bacterial endocarditis with consecutive emboli or small petechial hemorrhages

The red and raised nonhemorrhagic Osler nodes and the tender fingers and toes often described in adults are seldom seen in children

Anemia was present frequently in our patients all of them had hemoglobin levels below 12 gm and some below 8 gm Whether the anemia is the result of interference with iron metabolism such as is assumed to be the case in many infections or whether a specific hemolytic or "toxic" effect is involved is not definitely known

Two of our patients did not have positive blood cultures in both of these postmortem confirmation of the disease was obtained In at least half of our patients the blood cultures were reported as positive only several days after the onset of treatment Elevated sedimentation rate is

a nonspecific manifestation of subacute or chronic infection The high leukocyte count in nine of the twelve cases was caused principally by an increase in polymorphonuclear leukocytes but in three cases monocytes dominated the picture Macroscopic hematuria was not present in any instance microscopic hematuria was found in four cases The electro cardiographic and x ray changes were dependent on the underlying heart disease

Differential Diagnosis

In trying to establish the diagnosis of subacute bacterial endocarditis rheumatic fever should first be excluded Among the major manifestations of rheumatic fever only carditis may be present in subacute bacterial endocarditis On the other hand a number of the minor manifestations (fever abdominal pain precordial pain rashes epistaxis and similar laboratory findings) may frequently be found in both diseases

A positive blood culture makes mandatory the diagnosis of subacute bacterial endocarditis the presence of chorea polyarthritis or subcutaneous nodules favors a diagnosis of rheumatic fever Another clinical point

Table 11 Practical Differential Points between Subacute Bacterial Endocarditis and Active Rheumatic Fever

	Active R F	S B E
Polyarthritis	+	0
Nodules	+	0
Chorea	+	0
Erythema marginatum	+	0
Pericarditis	+	0
High antistreptolysin titer	+	0
Emboli	0	+
Spleen	0	+
Positive blood culture	0	+

may be helpful namely pericarditis almost never occurs in patients with subacute bacterial endocarditis whereas it is commonly associated with the severe carditis of rheumatic fever High antistreptolysin titers are almost never present in subacute bacterial endocarditis on the other hand severe anemia is a fairly common finding The principal points in the differential diagnosis between subacute bacterial endocarditis and rheumatic fever are summarized in Table 11

Almost all the systemic and localized infections occurring in patients with heart disease should also be considered in the differential diagnosis especially the enteric diseases pyelonephritis pneumonias

Course and Prognosis

Before the introduction of sulfonamides subacute bacterial endocarditis was practically always fatal Lichtman in 1943 [11] placed the

spontaneous recovery rate at about 1 per cent The same author found a 5 per cent recovery rate when sulfonamides were used in the treatment Today because of penicillin and other potent antibiotics the recovery rate is not less than 65 per cent In our series of twelve children with this disease in the past ten years all but two survived

Apparently the success of treatment depends a great deal on how early the disease is recognized According to Friedberg [1,2] if intensive therapy is started within a month after the onset of illness the recovery rate is 90 per cent or better whereas if the condition goes unrecognized for three months or more as many as 50 per cent of the patients die

Another factor to be taken into consideration is the age of the patient older people are less likely to recover than younger ones The isolation of a microorganism from the blood stream enhances the success of treatment considerably Whether the poor prognosis for patients with negative blood cultures is due to the delay in treatment caused by a search for the etiologic agent or whether the same set of circumstances that prohibits the bacteria from getting into the circulation prevents antibiotics from penetrating the vegetation is not known at present It is a fact however that patients with negative blood cultures have a poorer prognosis Obviously the more sensitive the isolated organism is to the commonly used antibiotics the more successful the treatment will be

TREATMENT

In view of these factors there has been a tendency within the past five years to start treatment of patients suspected of having subacute bacterial endocarditis before the causative organisms have been identified This policy is predicated not only on the advantages of early therapy but also on the unlikelihood of obtaining a positive blood culture after the first few hospital days Finland[119] stated that "from the bacteriologic point of view however a delay of more than forty eight hours or at most five days is logistically unsound and does not seem justified provided the treatment is begun only after adequate samples of blood for culture have been obtained during this waiting period for later examination" The chance of obtaining a positive blood culture later in the disease if it was impossible during the first five days is practically nil As a matter of fact most of the positive cultures are obtained within the first forty eight hours

Our policy within the past few years then has been one of suspecting subacute bacterial endocarditis in all children with known heart disease who have fever of unknown origin of at least one week s duration If in addition to fever one or more of the following criteria are present the diagnosis of subacute bacterial endocarditis is made clinically (1) splenomegaly (2) embolic phenomena and (3) clubbing in a patient without arterial unsaturation Several blood cultures (at least six) are obtained within the first forty eight hours especially during chills or

sudden elevation of temperature In cases of coarctation of the aorta, arterial culture is also obtained At the end of a two to five day waiting period during which all pertinent differential diagnostic possibilities are explored penicillin treatment is begun even if the cultures are not yet positive

The drug of choice depends on the availability and the sensitivity of the organism Since the alpha streptococcus is the commonest etiologic agent penicillin alone or in combination with streptomycin is the most commonly used therapeutic agent The dosage of penicillin should be between 3 and 10 million units per day given either in a continuous intravenous drip or by the intramuscular route in six divided doses To begin with these patients are usually given 3 to 6 million units of penicillin per day and only if this does not seem to achieve the desired drop in temperature and sterilization of the blood stream within a week do we switch to a higher dosage schedule Streptomycin in doses of 0 25 to 0 50 gm every eight hours may also be added at this time On the basis of our own experience and of the theoretic considerations outlined by Finland[119] we believe that aqueous solutions of sodium or potassium salt of penicillin should be used rather than the longer acting repository form at least for the first four weeks Probenecid a renal excretory inhibitor of penicillin may be utilized to elevate the level of penicillin in the blood 1 or 2 gm per day in four divided doses can be given by mouth The use of this agent in children is limited

Instead of penicillin or streptomycin other drugs such as bacitracin the tetracyclines chloramphenicol erythromycin or polymyxin may be used in cases in which the organism seems sensitive only to one of these agents However *more than 90 per cent of the cases of subacute bacterial endocarditis are caused by* Streptococcus viridans *an organism extremely sensitive to adequate doses of penicillin* Whatever drug is used it should be continued for at least six weeks Recent reports in the literature suggest the combined use of penicillin and streptomycin for two weeks only however I consider the evidence in favor of such a short cure insufficient for the time being It may be permissible though if the patient s condition progresses satisfactorily to substitute one daily injection of a repository penicillin in comparable doses for the last two weeks of treatment

One of the most disturbing features in the treatment of subacute bacterial endocarditis is the persistence of a low grade fever without positive blood cultures or any other signs of activity In our experience this is commonly due to the local irritant effect of a large number of penicillin shots given intramuscularly to a child with a greatly wasted musculature I feel that only with the utmost nursing skill the use of the sharpest needles and a carefully mapped out rotation of the injection sites can this be avoided Even with the best of nursing care local irritant action may be unavoidable sometimes only the substitution of

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developing after dental extractions has been demonstrated by Palmer and Kempf [64] who found transiently positive blood cultures in fourteen of eighty two patients after dental extraction

Depot penicillin (600 000 units) is recommended on the day of and the day after dental extraction or other type of oral surgery (tonsillectomy and adenoidectomy) Febrile illnesses of any sort in patients having congenital or rheumatic heart disease should be treated by some antibacterial agent for at least one week

Although liberal chemoprophylaxis and chemotherapy is probably extremely useful in preventing the development of subacute bacterial endocarditis it must be pointed out that it is not uniformly successful and that it may not be without danger One must bear in mind the development of resistant strains the occurrence of sensitivities and a change in the bacterial flora

COMPLICATIONS

Since the introduction of effective agents against organisms causing subacute bacterial endocarditis it has become obvious that some patients may not recover in spite of a bacteriologic cure In a considerable proportion of these death is caused by congestive failure at postmortem examination no organisms can be cultured but evidences of severe myocardial damage are found According to our experience this does not seem to represent a serious risk in children all the patients in whom we were able to sterilize the blood stream recovered satisfactorily

In one of our patients however—a boy with true truncus arteriosus who recovered from subacute bacterial endocarditis—polyarthritis pericarditis and a typical picture of acute rheumatic carditis developed within two months after his blood cultures had become sterile He responded well to treatment with ACTH Whether this represented a complication of subacute bacterial endocarditis or whether it was in fact true rheumatic carditis cannot be stated with certainty It is simply mentioned as an interesting and therapeutically approachable condition developing in the wake of bacteriologically cured subacute bacterial endocarditis

ACUTE BACTERIAL ENDOCARDITIS

INCIDENCE AND ETIOLOGY

Septicemia with invasion of the endocardium is called acute bacterial endocarditis It is a rare condition today only about half a dozen such instances having been seen at The Children s Medical Center within the past ten years

Acute bacterial endocarditis in contrast to subacute bacterial endocarditis should terminate favorably or unfavorably within two months Also whereas the cardiovascular phenomena dominate the picture in

subacute bacterial endocarditis in acute bacterial endocarditis these are overshadowed by the general picture of sepsis No single organism is implicated in the majority of cases unlike subacute bacterial endocarditis in which the alpha streptococcus is responsible *Staphylococcus aureus beta hemolytic streptococci pneumococci and Escherichia coli* are the microorganisms most frequently encountered in this condition in children However any other organism causing generalized blood stream infection may attack the endocardium

Pathology

In acute bacterial endocarditis the picture is dominated by thrombotic masses (*vegetations*) consisting of fibrin and bacteria which invade the valvular or parietal endocardium Pre existing heart disease is not a prerequisite as it is in patients with subacute bacterial endocarditis Further pathologic differences between the two conditions are the much more severe destruction ulceration perforation (endocarditis ulcerosa) and the purulent or serosanguineous pericarditis commonly present with acute bacterial endocarditis

Any and all organs may be invaded by the septic process Splenomegaly is the rule Pneumonitis with abscess formation is common The kidneys are involved in a high percentage of cases they may show *multiple abscesses parenchymal or perinephric and even evidences of* glomerulonephritis or infarct Meningitis and meningoencephalitis may be present as well

Clinical Picture

Within the framework of generalized sepsis there may be only a few signs pointing to the involvement of the cardiovascular system The *most clear diagnostic finding is the appearance of a loud heart murmur* in a patient with septicemia Other less pathognomonic findings are the appearance of progressive cardiac enlargement and congestive failure and the presence of embolic phenomena Janeway spots—painless hemorrhagic lesions on the palms or soles—are also characteristic of acute bacterial endocarditis

Diagnosis

The diagnosis is almost solely of academic importance It should be possible to make the diagnosis of a blood stream infection with ease if the signs mentioned previously are present a localization of the septic process in the endocardium must be suspected

Treatment

Whether *or not there is cardiac involvement* the treatment of septicemia should be the same it consists in adequate antibacterial treatment with penicillin streptomycin or any other agent that proves

to be effective according to sensitivity tests The dosage schedule outlined for penicillin and streptomycin in subacute bacterial endocarditis should be adequate in instances of acute bacterial endocarditis

There are no mortality figures based on large series available in the pediatric literature In 1947 Wilhelm and his associates[3, 6] analyzed forty cases of acute bacterial endocarditis in adults a mortality of 60 to 80 per cent was noted depending on the causative organisms Two of our six patients died one with *Escherichia coli* endocarditis and one with a staphylococcus infection The other four had *Staphylococcus aureus* septicemia

CHAPTER 9

Arrhythmias

The *normal rhythm of the heart is dependent on sinoatrial impulses* discharged at regular intervals and spreading through the conduction system at conventional speeds The impulse travels rapidly from the sinus node through the atria and arrives at the atrioventricular node The passage through the atrioventricular nodal tissue is slow--about one fifth of the speed in the atria From the atrioventricular node the impulse spreads at an increased speed down the bundles of His stimulating the left side of the muscular septum first then it is conducted through the Purkinje fibers and through the ventricular wall proper from endocardial to epicardial direction (Figs 35 36 p 46)

In the healthy child this mechanism results in a regular rhythm at a rate of 80 to 180 beats per minute The actual rate in the normal person depends a great deal on his age and his state of mind On the whole the younger the child the more rapid is the pulse Rates as high as 180 per minute are occasionally encountered at birth though a sig nificant drop usually occurs in one or two hours time During the first week of life the average rate is less than 140 beats per minute and during the first year less than 120 By the time a child is six years of age the sleeping pulse rate is usually less than 100 beats per minute and at puberty *as a rule less than 80 Obviously all these are average values* and are subject to a great deal of fluctuation through crying excitement fear and so forth Consequently *pulse rates* to mean anything at all *must be taken during sleep* especially in children

Changes in this regular rhythm at average rate are called arrhyth mias The arrhythmias will be discussed according to the following classification (*a*) arrhythmias with sinus pacemaker (*b*) arrhythmias with ectopic pacemakers and (*c*) arrhythmias caused by conduction defects

ARRHYTHMIAS WITH SINUS PACEMAKER

SINUS TACHYCARDIA

Sinus tachycardia is the rule rather than the exception in children being examined by a doctor most youngsters exhibit higher than normal pulse rates because of anxiety when they enter a doctor s office Exercise fever anemia or shock may also cause an increased pulse rate Rheu

matic carditis is accompanied by a persistently elevated heart rate even in sleep which is out of proportion to the accompanying temperature Congenital heart disease without cardiac insufficiency does not usually give rise to tachycardia Thyrotoxicosis a common cause of sinus tachy cardia in adults is rare in children occasionally it may be encountered in girls at the age of puberty

Clinically sinus tachycardia is suspected if the heart rate is between 140 and 200 beats per minute and does not have a clocklike regularity and consistency If one spends a few minutes listening to the heart or counting the radial pulse minor changes of 10 to 15 beats per minute will frequently be observed This is in marked contrast to the patient with paroxysmal atrial tachycardia whose heart rate is rigidly fixed and is almost always more than 180 beats per minute Reflex vagal stimu lation (eyeball pressure carotid sinus stimulation or Valsalva man euver) often causes slight but definite alterations in the heart rate of patients with sinus tachycardia.

Electrocardiographically sinus tachycardia can easily be recognized by the *well formed P waves a P R interval of 0 1 to 0 2 second* and a normal appearing QRS complex (Fig 70 p 76)

In most instances sinus tachycardia does not require any specific treatment—only search for and elimination of the cause (shock or ane mia) Digitalis has no influence on sinus tachycardia without underlying heart disease Phenobarbital sometimes slows the pulse of a hyperex citable youngster

Sinus Bradycardia

Sinus bradycardia in adults is commonly defined as a heart rate of less than 60 beats per minute It is obvious from the previous discussion of the normal heart rate of healthy children that no such clear cut defi nition is possible in the field of pediatrics By and large rates of less than 100 in infants and 80 in children should be considered bradycardic

Sinus bradycardia is rare in healthy children but is seen occasion ally in athletes and in patients with rheumatic fever increased intra cranial pressure increased systemic blood pressure typhoid fever or hypothyroidism The appearance of atropine resistant sinus bradycardia during cardiac surgery is an ominous sign which may indicate profound anoxia leading gradually to cardiac standstill and death Electrolyte disturbances especially potassium intoxication may also be accompanied by profound bradycardia

The clinical differentiation of sinus bradycardia from a slow pulse rate resulting from nodal rhythm is impossible On the other hand in complete heart block a fairly common cause of bradycardia careful observation of the jugular venous pulse reveals atrial pulsations (A waves) during ventricular diastole On scrupulous auscultation the in

tensity of the first heart sound at the apex shows definite changes in patients with atrioventricular dissociation (see p 206)

In most instances the electrocardiogram is essential in making a definite diagnosis of sinus bradycardia The tracing is characterized by a normal P wave a P R interval of less than 0 20 second and a normal QRS complex (Fig 71 p 76)

Sinus bradycardia in children does not require symptomatic treatment careful search for the underlying cause should be made however

Sinus Arrhythmia

Sinus arrhythmia is a phasic change in the rate of discharge of sinus impulses resulting in a rhythmic speeding and slowing of the heart rate The pulse rate increases with inspiration and decreases with expiration The presence of sinus arrhythmia is predicated on increased vagal tone and therefore is almost never present in young infants but is pronounced in adolescents Slow rates predispose to sinus arrhythmia and fast rates abolish it By and large sinus arrhythmia is a sign of good cardiac reserve although it has been noted with congenital as well as rheumatic heart disease

The diagnosis is easily made without the use of the electrocardiograph The pathognomonic feature is a phasic variation of the heart rate synchronous with respiration It can readily be abolished by the tachycardia caused by exercise

Electrocardiographically the QRS complexes and the P R intervals are regular and unchanging whereas the P P and R R intervals are different at various portions of the tracing (Fig 72 p 76)

No treatment is indicated

ARRHYTHMIAS WITH ECTOPIC PACEMAKERS

All parts of the conduction system from the sinus node to the Purkinje fibers have basic impulse forming properties Under ordinary circumstances the sinus node because it discharges impulses most rapidly is the pacemaker of the heart all other potential pacemakers have an inherently slower discharge rate hence they do not usually have an opportunity to take over the lead

Under certain circumstances however such as decreased irritability of the sinus node (sinus bradycardia or marked sinus arrhythmia) blockage of sinus impulses (sinoatrial block) or acquired hyperirritability of one of the lower centers the sinus node may lose control of the rhythm of the heart Under these conditions one of the ectopic pacemakers in the atria in the atrioventricular node or in the ventricle proper may become the governing body of the heart temporarily or permanently If the reign of the ectopic pacemaker lasts for one or two beats only ectopic or premature beats occur If the sinus node re

linquishes its control for several cycles in succession ectopic tachycardia results.

Ectopic Beats

Ectopic beats appear to be less common in children than in adults but no recent figures on their incidence are available Lyon and Rauh[229] found ectopic beats in 0 06 per cent of a large series of healthy newborns and in 2.2 per cent of healthy school children Landtman[198] in his detailed book on arrhythmias of children found ectopic beats in 1.53 per cent of his cases Supraventricular premature beats are more common in children than ventricular ectopic beats

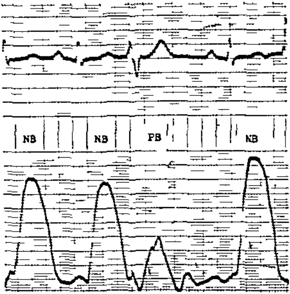


Figure 135 Electrocardiogram (top) and right ventricular pressure tracing obtained with cardiac catheterization (bottom) The first electrocardiographic beat is followed by a normal beat in the right ventricle The second QRS complex is equally followed by a normal ventricular complex. The third QRS complex is a premature one and is followed by a low pressure tracing within the ventricle After this a compensatory pause follows, and the normal beat following the compensatory pause is considerably higher in the right ventricle than the normal beat and the premature beat

Physiologically the premature beat is a less than completely effective contraction because of the shorter than normal diastolic filling of the ventricle In contrast, the succeeding normal beat, if separated from the ectopic one by a compensatory pause is a stronger than average contraction because of the extra long diastolic filling time (Fig 135)

Etiology

The majority of patients with ectopic beats have normal hearts However rheumatic fever subendocardial sclerosis, myocarditis and

drugs (digitalis quinidine) may all contribute to their occurrence Fortunately for the pediatrician coronary disease pregnancy alcohol and tobacco—all common causes of premature beats in adults—do not have to be considered in the etiology of this arrhythmia in the majority of children

Clinical Picture and Differential Diagnosis

Premature beats usually cause no symptoms although older children may find them annoying if they occur several times per minute It may be of interest to mention that the extra strong postextrasystolic beat is often noted by the patient as a thump whereas the premature beat itself may pass by quite unobserved Supraventricular that is atrial or nodal premature beats should be suspected from the clinical examination if less than a complete compensatory pause is present In contrast ventricular premature beats are followed by a complete compensatory pause One further point in differential diagnosis is that whereas the first sound of the supraventricular premature beat is always accentuated the first sound of the ventricular premature beat varies from normal to loud to diminished [85] depending on the time interval between the QRS complex and the preceding P wave If the atrial contraction occurs only a short time (less than 0 12 second) before the QRS the first sound will be loud If the P R interval is long the first sound will be dull

Ectopic beats originating from a normal heart are usually easy to distinguish from those occurring in a diseased heart To begin with premature beats are seldom if ever the sole manifestation of heart disease in children if carefully searched for other evidences of cardiac abnormality (murmur cardiomegaly electrocardiographic evidence of hypertrophy or elevated sedimentation rate) are demonstrable if there is underlying heart disease Exercise abolishes premature beats originating from a normal heart, whereas they are commonly exaggerated on exercise if heart disease is present The presence of heart disease should be suspected if the premature beats show varying configuration (suggesting multifocal origin) in the electrocardiogram On the whole ventricular premature beats are relatively more common in patients with heart disease than in those whose cardiovascular system is intact

Excess digitalis administration usually causes supraventricular premature beats in children although the occurrence of ventricular premature beats particularly in bigeminal rhythms is practically diagnostic of digitoxicity Quinidine intoxication is more likely to cause ventricular than supraventricular premature beats

A series of ectopic beats (usually though not invariably supraventricular in origin) may be encountered in newborn babies Keen obstetricians have discovered the presence of this arrhythmia when listening to the fetal heart The ectopic beats may be so numerous that the rhythm is totally irregular but the over all rate is seldom more than 140 to 160

beats per minute The prognosis of this "arrhythmia of the newborn" is usually good and if not accompanied by other evidences of heart disease it usually disappears without treatment within the first ten days of life It should be electrocardiographically differentiated however from congenital atrial flutter a rare and serious abnormality usually associated with subendocardial sclerosis

The diagnosis of premature beats should usually be based on the electrocardiogram Marked sinus arrhythmia sinoatrial block and occasionally even atrial flutter or fibrillation may have to be considered in the differential diagnosis Although exercise tests inspection of the jugular venous pulse and careful observation of the heart sounds may aid in determining the nature of the arrhythmia by clinical means in most instances the little effort and expense spent on the electrocardiogram in return for an accurate authoritative explanation of the child's condition are fully appreciated by the parents

Treatment

Premature beats in themselves unless they occur frequently require no treatment but the question of underlying heart disease should be given careful therapeutic consideration in every instance Ventricular premature beats are always more worrisome than supraventricular ones because of their possible extension into ventricular tachycardia

If the ectopic beats are clearly the result of psychological factors in an unstable adolescent reassurance and the judicious use of phenobarbital or Serpasil may abolish them If treatment by these simple methods is not successful and the arrhythmia is of concern to the patient then specific cardiac drugs may be administered depending on the site of the pacemaker Supraventricular premature beats should be treated with digitalis ventricular premature beats with quinidine or Pronestyl Potassium chloride administered orally may be effective for both supraventricular and ventricular premature beats The dosages are outlined in Table 12 Before either digitalis or quinidine is administered the physician must be certain that these drugs do not in fact cause the arrhythmia If any doubt exists it is a good policy to discontinue all medication for a period of days or weeks

Ectopic Tachycardia

A series of premature beats at rapid rates is called ectopic tachycardia The duration of the ectopic rhythm may be from a few seconds to several years

Simple Paroxysmal Tachycardia

Usurpation of the normal sinus rhythm by an ectopic focus results in simple paroxysmal tachycardia characteristically the paroxysm begins and ends suddenly much as one would abruptly shift the gear of an

Table 12 Principal Drugs Used in Pediatric Cardiology

D g	I t f Adn t t	D		T ty	I d cat ns
		In t l	Ma te		
A t g tiv D git in (D gital N tl ll)	P O I M	<2 yrs 0 02–0 03 mg /lb >2 yrs 0 01–0 0 mg lb	1/10 ı ıt l do c d ly	N us a miting a rhythm as p l g d P R	Co gesti e f lure p xysm l atr l ta hycardia
D g x n	P O	<2 y s 0 03 0 04 mg /lb > y 0 0 –0 03 mg /lb	1/4 ı ıtıal dos daily	S mılar to digıtoxın	C ngestiv fa lu e pa o ysmal atr al tachycardı
	I a e t r l	<2 yrs 0 02 0 03 mg /lb > yrs 0 01–0 0 mg /lb	1/5 ı ıtı l dose da ly		
La t s d C (Ced l nid)	I M o I V	<2 0 02–0 03 mg /lb >2 yrs 0 01–0 0? mg /l	1/3 1/2 n t al d se d ıly	S mil r to d git x n	S me
M r uhydrin	I M	0 1 1	Same as initıal	C mp low s d m syn dr me acidosı dehyd atı	C gest e fa l e
Thi m n (M rc pt meri)	S C o I V	0 1 1 cc	S me a ın t l	S m as M r uhyd n	Co g t ve f ıl
Ami phyll ne	I V r I M	0 1–0.2 gm		H daches p lp tati n l w blood pr ssur	Dıu tic alo e or ppo t ng m r u y B onchod lato
Amm nium hl rid	P O		0 5 gm X 2 3 da ly	Acidosıs GI upset	Dıu t supp rting m r u y
Di m	P O		50 2 0 mg /day	No e	Dıu t alo e o supp t g me cu y
Mo ph ne	S C	1 mg /10 lbs	q 4 ho rs	CNS d pressi respı at ry f l e v mitı g	Ano i sp lls x item nt w th a rhythmı p lmo ry d m
A t hythmic Qu di	P O	1 2 3–400 mg q 2 X 8 r 1 3–4 500 mg q 1 in s cess d es	Effecti dose ry 4–6	N usea v miti g tin tus wid ned QRS c mpl es ven t i ul r a rhythm as	Ve t i ula a hythmia At ıal fib ill tio Atrı l flutte

P ca m d P y	I O I M I V	7 mg b 50–300 mg 100 n g	50 300 mg q 6 Effec dos q 6 R p q 5 10 m u es	Hyp V u hy hm as	V n a hyth as
Neos m P g	S C	0 05 0 5 mg		I yp y f GI mus- h g d hyp	P y ma n l ta hy d
Eph dn	P O I M	1 7 mg 10 b	q 6 h urs	S m t p ph	S w d ri la hy hm
Ep ph	I M I a d	0 1 0 3 f 1 1000		T hy hyp ns ess ess	C d c es I yp n
C h d	I V I d	2 4 f 10ᵉ s	R p y 30 t 60	C d d p especi y d p ts	S w d weak d
A hyp R w fi p S p	P O		0 1 0 2 mg 3 × d y	D w ese as ges d h	Iyp ns A hythm
I yd Ap es	I O I M	10 50 mg 0 1 mg b	4 × d y q 4 h rs	I p h d h s	Hyp te n
P	P O I V	0 5 1 0 mg 0 5 1 0 g /lb	R p t 4 × d y R p t e y 30 m es	I yp ns use v g	Hyp n
A g m	I M I V	25 o 0 1 cc b 3 s 0 5 100 mg b e 1 p d	R p a y 4 6 I p ssa y	P y p y f b dyca h n	Hyp ens
H m h m d	P O S C I V		2 75 mg 4 × d y 0 1 1 0 n g kg	C p ur y p hyp	S hyp
A h p s Fh ph hyd h d N o- y ph	I V	0 1 0 5 mg s		B dy d h p y hy hm	Hyp p y m l a a hy d a
N p ph Le ph d	I V	4 g m 5ᶜ d se Tes d f 1 μg 10 kg h d us		B dy d hype y a hy hm	I yp ho k

automobile from a lower to a higher speed. Paroxysmal tachycardia is not a common condition in children. At The Children's Medical Center in Boston the yearly admission rate of new patients with paroxysmal tachycardia is about two. The average pediatrician may see one such case every five years or so. Nevertheless it may be worth while to spend considerable time in discussing this arrhythmia, since it represents one of the real treatable cardiac emergencies in children.

For obvious reasons there are not many physiologic studies of people with paroxysmal tachycardia. A brave soul would he be who would attempt cardiac catheterization under these conditions. A few studies are available, however, indicating considerable increase in the arteriovenous oxygen difference and some decrease in the arterial saturation during a paroxysm.[97] Animal experiments point to a decrease in the coronary blood flow and the mean arterial pressure during the attack. These changes are more pronounced with higher rates. Ventricular tachycardia results in a greater change in the coronary circulation than supraventricular tachycardia at a comparable rate.[349]

Clinical Picture

Paroxysmal rapid heart action may be supraventricular or ventricular in origin, and a clinical differentiation of the two types is often possible. The rate of supraventricular tachycardia is perfectly steady from one minute to the next, whereas ventricular tachycardia may show minor variations in rate. Reflex vagal stimulation (eyeball pressure, carotid sinus stimulation, Valsalva maneuver) may influence supraventricular tachycardia, whereas it has no demonstrable effect on ventricular tachycardia. At auscultation the intensity of the first heart sound is increased but unchanging in supraventricular tachycardia, whereas recurrent changes in intensity are often noted in ventricular tachycardia.

A clinical differentiation between the two principal types of supraventricular tachycardia, that is, atrial flutter and atrial tachycardia, is difficult. Reflex vagal stimulation, if effective at all, converts atrial tachycardia into normal rate and rhythm, whereas its effect on atrial flutter is only one of altering the atrioventricular conduction, resulting in an arithmetical division of the rate, such as halving. Exercise, by the same token, does not change the rate of paroxysmal atrial tachycardia at all, whereas in paroxysmal flutter it may cause the rate to increase in a mathematical ratio by one third or one half through changes in atrioventricular conduction.

The following clinical profile of simple paroxysmal tachycardia is based on our observations of forty-one cases seen at The Children's Medical Center during a twenty-year period.

The etiology of paroxysmal tachycardia often is not known. Slightly

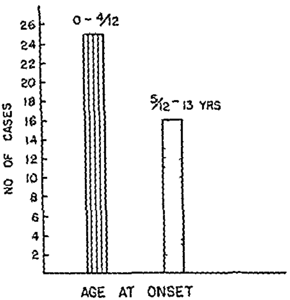


Figure 136 Age at onset in forty one patients with paroxysmal tachycardia [4]

more than 10 per cent of our patients showed electrocardiographic evidences of the Wolff Parkinson White syndrome another 20 per cent had congenital heart disease About 20 per cent of the patients had infections injuries or heart tumors or were under the influence of drugs at the time tachycardia occurred Whether or not any of these findings had anything to do with the occurrence of paroxysmal tachycardia would be hard to prove but in 50 per cent of the patients not even such circumstantial evidence was available to explain the occurrence of paroxysmal rapid heart action

Analysis of the age at onset in our forty one patients resulted in the interesting observation that the disease was more likely to start within the first four months of life than throughout the rest of childhood (Fig 136) It was also found that in most cases occurring in young infants no etiologic factors could be detected Conversely in older infants and children factors were frequently present that might explain the occurrence of the tachycardia Finally when the etiologic factors were correlated with the sex incidence it was learned that most patients with "idiopathic paroxysmal tachycardia were males (Fig 137) Thus on the basis of age sex and the presence or absence of possible etiologic factors paroxysmal tachycardia in children divides itself into two groups one consisting of males less than four months of age without concomitant disease and the other of older infants and children of either sex who may or may not have demonstrable etiology for tachycardia

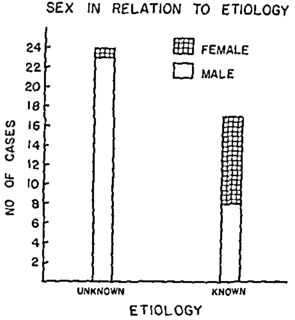


Figure 13. Sex in relation to etiology of paroxysmal tachycardia in children [2,6]

Seven of our forty one patients were not sick and their paroxysmal tachycardia was discovered during a routine physical examination The majority of the infants however were very ill indeed the picture resembling severe pneumonia or septicemia These babies were irritable and restless with ashen gray or cyanotic color and cold clammy skin Respirations were rapid and labored and often a hacking cough was present Cardiac enlargement was manifest and the heart rate was rapid even uncountable with a thready pulse Rales were heard over the chest and the liver was enlarged Peripheral edema and puffiness of the face were noted in some

Fever was noted occasionally In a few instances it could be explained on the basis of infection alone in others it was assumed to be due to congestive failure

A systolic murmur was present during the attack in almost half of the patients but in only a quarter of this group—all with murmurs of at least grade 3 intensity—did the murmur persist after cessation of the attack No diastolic murmurs were observed

Results of laboratory examinations were usually not of much assistance Leukocytosis if present could be explained by infection or congestion

The radiologic findings were those of congestive failure with "non specific" cardiac enlargement of varying degree and pulmonary conges

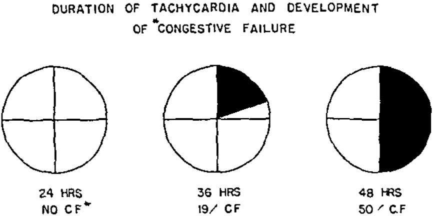


Figure 138 Relation of the duration of tachycardia to the development of congestive failure[256]

tion Isolated left atrial prominence was occasionally demonstrable The cardiac beat under the fluoroscope was rapid and feeble

Congestive failure developed in slightly more than 50 per cent of our patients with paroxysmal tachycardia most of the signs and symptoms described are manifestations of combined left and right ventricular failure Analysis of our cases revealed that three factors seemed to influence the development of heart failure (1) the rate—failure did not develop in any patient with paroxysmal tachycardia in whom the rate was less than 180 beats per minute (2) the duration of the tachycardia (Fig 138)—the longer the paroxysm lasted the more likely it was to *lead into congestive failure (3) the age of the patient—the younger the* child the more likely was congestive failure to develop perhaps because the manifestation of cardiac failure was the first symptom noted by the mother of a baby too young to complain

The electrocardiograph is the only consistently reliable tool for the accurate analysis of these arrhythmias About 95 per cent of our cases fell into the category of supraventricular tachycardia that is paroxysmal atrial tachycardia (Fig 76 p 78) or paroxysmal flutter Only 5 per cent of the patients showed characteristics of ventricular tachycardia (Fig 83 p 83)

The heart rate during the attack varied between 160 and 330 beats per minute Patients with rates of 300 or more almost always exhibited flutter (f) waves in leads II and III P waves were rarely identifiable they were either absent or superimposed on the preceding T wave If P waves were present the P R intervals varied from 0 08 to 0 13 second In practically all instances of supraventricular tachycardia the QRS complexes were normal in configuration and similar to the ventricular

complexes between attacks Occasionally however because of the presence of intraventricular conduction block abnormal QRS complexes resembling ventricular tachycardia may be present in patients who have in fact paroxysmal atrial tachycardia In these cases exact determination of the P QRS relationship or observation of the effect of vagal stimulation helps to identify the type of tachycardia accurately The QRS complexes in ventricular tachycardia are by definition bizarre and wide and unlike the complexes seen between attacks The T waves as mentioned usually blend with the preceding atrial waves hence it is difficult to describe their characteristics

After cessation of the paroxysm the majority of the children in our series showed perfectly normal electrocardiograms for their age In some evidences of left and right atrial hypertrophy persisted for a few days In others electrocardiograms characteristic of the Wolff Parkinson White syndrome were found In a few short P R intervals without corresponding widening of the QRS complex were observed Diphasic or inverted T waves in leads I and II with depressed S T segments were noted in some cases immediately after cessation of severe tachycardia of long duration these changes much like the P wave abnormalities were transient and lasted for a few days only We believe that the P wave changes are due to increased atrial pressure caused by congestive failure the T wave abnormalities may be an expression of myocardial ischemia

Treatment

Simple paroxysmal tachycardia can be successfully treated in almost every instance Before treatment is begun it is extremely important to ascertain whether the tachycardia is supraventricular or ventricular in origin since the therapeutic approaches to the two groups are entirely different

The treatment of supraventricular tachycardia should begin with reflex vagal stimulation First carotid sinus pressure should be tried by palpating the carotid artery as high up the neck as possible and pressing it against the vertebral column first the right side then the left should be pressed never both sides at the same time The pressure should be sustained for about thirty to sixty seconds with the stethoscope on the chest and released immediately when the rhythm changes If this maneuver is not effective eyeball pressure should be attempted The pressure should be applied on the closed lids just below the supraorbital ridge one side at a time for about thirty seconds In older children if neither of these procedures is effective an effort may be made to stimulate the vagus nerve by asking the patient to perform a Valsalva maneuver or making him vomit by the administration of 1 or 2 teaspoonfuls of syrup of ipecac

In our experience reflex vagal stimulation by the methods described is usually successful for paroxysms of short duration without congestive failure However only rarely were we able to abolish the paroxysm in critically ill small babies by these methods

Drug therapy is the next step in the treatment of paroxysmal supraventricular tachycardia In our opinion digitalis is the drug of choice We have used digitoxin Digoxin and Cedilanid equally successfully All three of these digitalizing drugs may be given parenterally digitoxin and Digoxin may be administered orally as well For the appropriate digitalizing dose see Table 12 In most instances the calculated dosage can be given throughout an eighteen hour period in three divided doses The oral or intramuscular route is recommended for the average patient but in an emergency the intravenous route may be used safely If the calculated digitalizing dose does not convert the rhythm to normal more digitalis should be given cautiously about one sixth of the calculated amount may be administered every four hours until either the desired effect is achieved or evidences of toxicity appear Incidentally carotid sinus stimulation may be worth attempting after full digitalization it may be effective under these circumstances even if it had no effect before digitalization

Digitalis if given in appropriate doses has been successful in the treatment of all but one of our patients with paroxysmal supraventricular tachycardia during the past twenty years Only one person did not respond to digitalis but she responded easily to Prostigmin administered subcutaneously after full digitalization

Our experience in the treatment of paroxysmal atrial tachycardia with other drugs is limited Quinidine and procaine amide are recommended by some authors but in our experience their principal use is in the treatment of paroxysmal ventricular tachycardia Mecholyl has also been recommended for the treatment of this condition our experience with this drug is limited but rather unfavorable

Continuous electrocardiographic tracings obtained during postoperative development of paroxysmal atrial tachycardia have revealed the electrocardiographic patterns of conversion in a few instances They are frightening electrocardiograms indeed consisting of prolonged asystoles appearance of atrial flutter with block later supraventricular or ventricular premature beats and finally appearance of normal sinus rhythm Hellerstein and his associates observed these and other changes during conversion by means of carotid sinus pressure [13]

Ventricular tachycardia in contrast to supraventricular tachycardia does not respond to reflex vagal stimulation Treatment with digitalis is contraindicated in most instances though it has been successfully used on occasion when all other measures failed Quinidine or Pronestyl administered orally or parenterally are the drugs of choice in this arrhythmia

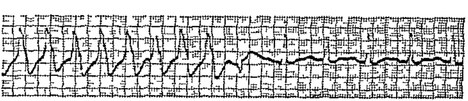


Figure 139 Successful use of Pronestyl in reverting ventricular tachycardia to normal sinus rhythm (lead II) Conversion occurred after the administration of 3 gm of Pronestyl given over a period of one hour Note (1) sudden reversion occurring within two beats (2) ventricular tachycardia at rate of approximately 140 before reversion (3) reversion to normal sinus rhythm with first degree heart block (pulse rate 0 28 second)

In our experience after a test dose of 25 to 50 mg 100 mg of quinidine every two hours should be given for eight doses If this does not work the next day 200 mg should be given every two hours again for eight doses The dose should be gradually increased in this way until either the tachycardia is abolished or toxic effects are noted Among the clinical signs of toxicity nausea tinnitus and vomiting should be mentioned The toxic effects on the electrocardiogram are manifested by widening of the QRS complex by more than 50 per cent of its original duration

Obviously if the patient is critically ill this leisurely schedule of oral administration of the drug cannot be adhered to In these instances quinidine should be given in progressively larger doses at hourly intervals starting with 100 mg and continuing up to 200 300 400 or even 500 mg until either therapeusis or toxicity is achieved In an emergency the drug may even be given intramuscularly

If quinidine is not effective in controlling ventricular tachycardia procaine amide given orally or intramuscularly should be tried The oral dose is 7 mg per pound of body weight, to be given every two to four hours Intramuscular administration for the sicker patient should start with 50 to 100 mg and should increase by increments of 50 mg every two hours until the arrhythmia is controlled or toxicity supervenes The principal toxic effects of procaine amide are hypotension or the development of multifocal ventricular premature beats In an emergency Pronestyl may be given intravenously under continuous electrocardiographic control At first 100 mg should be administered during a period of five minutes this should be followed by the same dose every five minutes until either the arrhythmia is controlled or toxicity occurs (Fig 139) This is a dangerous procedure and may produce ventricular fibrillation but if it is used cautiously it may serve as a lifesaving measure Seldom if ever should more than 1 0 gm be administered during one hour even for adolescent patients

Ventricular tachycardia caused by digitoxicity should be treated by withdrawal of the glycoside and administration of potassium chloride

Through the judicious and intelligent use of these rather simple tools most paroxysmal tachycardias in children may be reverted to normal rhythm without difficulty This of course is different from experience with adults The difference may be caused by the structurally intact heart underlying the vast majority of cases of paroxysmal rapid heart action in children One cannot emphasize often enough that if these children are treated rationally they should recover without difficulty and that there is no reason for drastic and hurried "catastrophe reactions when such a problem arises Of forty one patients with simple paroxysmal tachycardia treated at our hospital all but three recovered (One patient who died was treated with Mecholyl and one with an unusually high dose of digitalis accompanied by bilateral carotid sinus pressure) The third patient died from a cerebrovascular accident presumably caused by arterial embolism This complication with recovery has since been seen in one other child hence it may not be as infrequent an event as was thought previously

In addition to the efforts aimed at conversion of the ectopic rhythm to normal sinus rhythm the treatment of paroxysmal tachycardia should also consist in the administration of oxygen antibiotics and perhaps morphine for restlessness

The change in a desperately sick baby after the cessation of the tachycardia is truly remarkable Within a few hours the color returns the restlessness and cough disappear and the sensorium clears Within twenty four to forty eight hours the baby loses its edema occasionally with a weight loss amounting to as much as 10 per cent of the total body weight In a matter of days the liver returns to normal size and the transient post tachycardic P wave and T wave changes disappear Usually x ray examination shows clearing of the pulmonary congestion within a few days but the heart may not return to normal size for seven to ten days

Prophylaxis

The drug used to convert the rhythm to normal should be administered for at least a month for prophylactic purposes after which it *may be abruptly discontinued If recurrences are noted it should be* resumed for at least another six months At this point sudden cessation should again be attempted

Digitalis in maintenance doses is usually effective in preventing the recurrence of supraventricular tachycardia Patients with the Wolff Parkinson White syndrome should probably receive quinidine for prophylaxis in doses of 200 to 400 mg four times a day On occasion

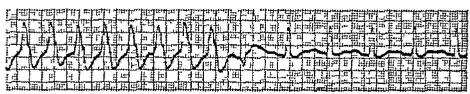


Figure 139 Successful use of Pronestyl in reverting ventricular tachycardia to normal sinus rhythm (lead II) Conversion occurred after the administration of 3 gm of Pronestyl given over a period of one hour Note (1) sudden reversion occurring within two beats (2) ventricular tachycardia at rate of approximately 140 before reversion (3) reversion to normal sinus rhythm with first degree heart block (pulse rate 0 28 second)

In our experience after a test dose of 25 to 50 mg 100 mg of quinidine every two hours should be given for eight doses If this does not work the next day 200 mg should be given every two hours again for eight doses The dose should be gradually increased in this way until either the tachycardia is abolished or toxic effects are noted Among the clinical signs of toxicity nausea tinnitus and vomiting should be mentioned The toxic effects on the electrocardiogram are manifested by widening of the QRS complex by more than 50 per cent of its original duration

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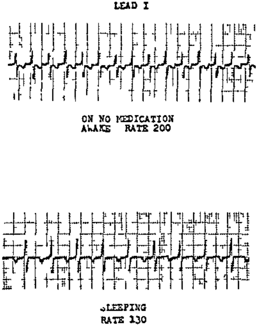


Figure 141 Effect of sleep on the rate of patient with ectopic atrial tachycardia

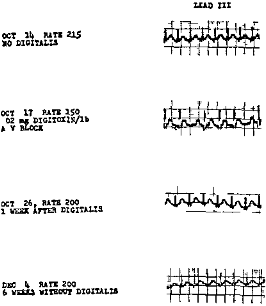


Figure 142 Effects of digitalis on ectopic atrial tachycardia Note the development of atrioventricular block and concomitant slowing of ventricular rate Note return of relatively normal atrioventricular conduction with rapid rate nine days after discontinuance of digitalis

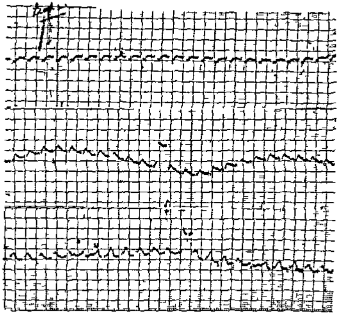


Figure 143 Ectopic tachycardia leads I II and III We have been following up this patient for seven years and her pacemaker has not changed although her rate has slowed down

One of these three cases corresponds to what Parkinson and Papp [96] called repetitive tachycardia (Fig 77 p 79) a transition between extrasystoles and paroxysmal tachycardia According to these authors in a long tracing an occasional premature beat may be seen alternating with short runs of tachycardia then again a few premature beats The ultimate prognosis in all Parkinson and Papps cases was good although in one instance the attacks lasted as long as ten years They recommended quinidine as the treatment of choice Our case occurred in an infant and lasted about three months but treatment with digitalis finally brought it under control with return of normal sinus rhythm

Our other two patients never had normal sinus rhythm while under our care Their P waves were always abnormal even at slow rates In the second case digitalis was able to reduce the ventricular rate by producing a second degree atrioventricular block (Fig 142) while a fast atrial rate persisted Serpasil in combination with digitalis slowed the pacemaker and maintained a 3 1 atrioventricular block thus resulting in the satisfactory ventricular rate of 80 to 100 beats per minute In the third patient (Fig 143) digitalis alone eventually slowed but did not change the atrial pacemaker Quinidine Pronestyl potassium and Prostigmin were not effective in these patients The rhythm in both these children today one at least seven years[4] and the other at least one and one half years after they were first seen is still under the control of an ectopic pacemaker Three cases of this sort have been reported recently all with good long term prognosis [162]

Paroxysmal Atrial Tachycardia with Block

A transitional stage between flutter and paroxysmal atrial tachycardia is an irregularity called paroxysmal atrial tachycardia with block. Electrocardiographically the arrhythmia consists of regular looking P waves at a rate of 140 to 180 and normal QRS complexes separated from the P waves by varying but always prolonged P R intervals,[-3] every fourth or fifth P wave is completely blocked (Fig 79 p 80). This arrhythmia is commonly present in patients with digitoxicity although we have seen it in children with primary myocardial disease who had never received digitalis and in whom the arrhythmia was abolished by administration of the glycoside.

Because the ventricular rate is only moderately rapid the arrhythmia itself probably never leads to congestive failure. However, reflecting the status of the myocardium or the presence of severe digitoxicity, paroxysmal tachycardia with block has serious prognostic implications. According to this line of thought the management of this arrhythmia should be directed principally toward establishing whether digitalis may have played a role in its causation. If this is the case, withdrawal of the glycoside and administration of potassium chloride is indicated. On the other hand, if the patient has not taken digitalis before, then cautious administration of a rapidly excreted glycoside such as Digoxin or Cedilanid, is indicated.

Atrial Flutter

One of the supraventricular arrhythmias most commonly associated with severe heart disease is atrial flutter. The atria here are in continuous regular activity at a rate of 200 to 350 beats per minute. The condition is rare even in adults (one in eighty patients with heart disease)[-35] and is much rarer in infants and children.

In its most benign form atrial flutter may occur in a previously normal heart and manifest itself as paroxysmal supraventricular tachycardia with a ventricular rate of 300 beats per minute or more. In this form it is no different from other types of simple paroxysmal tachycardia.

In contrast, atrial flutter accompanying rheumatic heart disease is of serious prognostic importance since in a child it indicates almost without exception the presence of active carditis. The association of flutter with congenital heart disease is of equally ominous significance, only the severest types of lesions, usually accompanied by congestive failure and gross atrial enlargement (large atrial septal defect, total pulmonary venous anomaly, ventricular septal defect with pulmonary hypertension) manifest this arrhythmia. Occasionally we have seen atrial flutter appearing transiently as a postoperative complication of closure of atrial septal defects. Finally we have encountered this arrhythmia in conjunction with subendocardial sclerosis in one fatal instance

this was the first manifestation of heart disease in a newborn infant and in another fatal case it was associated with digitalis intoxication

The electrocardiographic features of atrial flutter have been discussed and illustrated previously (see p 79 and Fig 49 p 57)

Two prevailing theories on the nature of impulse formation in flutter will be mentioned The older and in the eyes of many the best explanation for the rapid atrial activity is the one proposed by Lewis [08] Elaborating on theories of others he proposed that no one single focus is responsible for the presence of flutter but that the atria are in one continuous circus movement"—a wave of impulse incessantly going around the two atria In recent years Prinzmetal [7] using cinematographic techniques seriously challenged the validity of Lewis views and proposed his "unifocal" theory according to which the impulses in atrial flutter much as in paroxysmal tachycardia originate in one ectopic focus somewhere in the atria According to this theory of the atrial arrhythmias paroxysmal tachycardia and flutter are not qualitatively but only quantitatively different from each other

The proponents of this unifocal theory seem to increase in number with the years The only pertinent observation I can make on this subject also favoring Prinzmetal s theory is to point out the surprising ease with which paroxysmal tachycardia flutter and fibrillation may convert into each other within a matter of a few minutes or hours

Clinical Picture

The suspicion of atrial flutter should be raised in any patient who has a regular pulse rate of 180 beats per minute or more If by carotid sinus stimulation the rate can be decreased to one half or one third or if by exercise the rate can be increased in the same proportion the clinical diagnosis of flutter can be made with assurance Absence of these vagally induced changes does not however, exclude the diagnosis of atrial flutter

The clinical profile is not significantly different from that discussed in the section on Simple Paroxysmal Tachycardia except that because of the relatively frequent association of this condition with structural heart disease the symptoms may occur earlier and in more severe form

The effect of flutter on the circulation depends largely on the ventricular rate If only one third or one fourth of the impulses are carried to the ventricles the arrhythmia is tolerated relatively well although the patient feels better when converted to normal sinus rhythm If the ventricles respond in a 1 1 or 1 2 fashion congestive failure ensues

Treatment

The drug of choice in the treatment of flutter is digitalis One effect of the glycoside is to increase the atrioventricular conduction time and

by so doing to increase the ratio between atrial and ventricular beats by virtue of this ventricular slowing alone the patient's symptoms may improve a great deal and congestive failure may disappear Perhaps even more important digitalis may convert atrial flutter either directly into normal sinus rhythm or more commonly first into fibrillation and then to normal sinus rhythm Digitalis in the dosages indicated (Table 12 p 184) should be used

If flutter does not change into normal sinus rhythm after administration of an adequate amount of digitalis sudden cessation of the drug for two or three days may result in the desired effect If flutter or fibrillation still persists administration of digitalis in doses resulting in 1 3 or 1 4 block should be resumed and quinidine should be added to the regimen according to the schedule outlined on page 191 Quinidine should not be used in the treatment of flutter (or fibrillation) without previous digitalization since it may result transiently in a higher ventricular rate

Results of treatment vary with the underlying heart condition Almost all cases of flutter originating from a normal heart and manifesting themselves as paroxysmal atrial tachycardias can be successfully treated with digitalis Conversion in the patient with severe rheumatic or congenital heart disease cannot be expected in more than 50 per cent of the cases If atrial flutter persists in the face of rigorous attempts at conversion one ought to aim at maintaining a 1 3 or 1 4 atrioventricular block with digitalis

Embolic complications in conversion of ectopic tachycardia are rare in children although they have occurred at least twice in the last twenty years at The Children's Hospital

Atrial Fibrillation

The severest type of supraventricular arrhythmia is atrial fibrillation The atria are in continuous irregular motion at a rate of 400 to 700 beats per minute The ventricular response depending on the atrioventricular conduction is always only a fraction of the atrial rate but it is equally and totally irregular

Although idiopathic atrial fibrillation that is fibrillation originating from an anatomically normal" heart does reportedly exist in adults fibrillation in children always means structural heart disease usually severe active rheumatic carditis Severe congenital heart disease specifically atrial septal defect ventricular septal defect or Ebstein's anomaly of the tricuspid valve also may be associated occasionally with this irregularity On the whole it is not a common arrhythmia It occurs in less than 5 per cent of children with rheumatic heart disease[131] and considerably less frequently in patients with congenital heart disease

The electrocardiogram in fibrillation has been discussed and illustrated previously (p 81 and Figs 50 p 58 and 80 p 81) The type

for three minutes or more without cardiac massage Recovery, in a cardiac sense has been reported after considerably longer periods of ventricular fibrillation

Ventricular fibrillation cannot be diagnosed clinically but it may be suspected if a child during anesthesia operation or cardiac catheterization suddenly loses completely his blood pressure his peripheral pulses and heart sounds About 10 per cent of the patients with cardiac arrest have ventricular fibrillation

If one is fortunate enough to have an electrocardiogram connected to the patient one may follow these stages in ominous succession (1) occasional ventricular premature beats some with long Q T intervals (2) development of a series of ventricular premature beats into ventricular tachycardia (3) appearance of ventricular flutter—the QRS complexes of ventricular tachycardia widen until they appear as a continuous regular sine wave at a rate of about 200 beats per minute without a clear cut distinction of QRS complexes and T waves and (4) finally these complexes become irregular in size shape and rhythm This is called ventricular fibrillation

Treatment and Prevention

Prompt and well organized treatment of this condition is of utmost importance the patient s survival depends on action within sixty seconds The plan of treatment for cardiac arrest will be discussed later (see p 207)

Almost more important than the treatment of ventricular flutter and fibrillation is their prevention Since the occurrence of this often fatal arrhythmia in children is almost always dependent either on an operation cardiac or otherwise or on cardiac catheterization and not primarily on myocardial disease certain preventive measures are possible (1) Choose the proper anesthetic and method of induction (2) Avoid anoxia by the administration of an extra amount of oxygen at critical periods such as when dye is injected during angiography or when ventricular premature beats appear during operation (3) Prophylactic injection of procaine into the pericardial sac and into the myocardium if this is going to be incised Pronestyl should be administered intravenously if ventricular premature beats appear in any appreciable number (4) The electrocardiogram should be used continuously during operation to indicate the danger signals

ARRHYTHMIAS CAUSED BY CONDUCTION DEFECTS

Sinoatrial Block

The absence of a P wave caused by failure of the impulse to reach the atrium from the sinus node is called sinoatrial block Since the only indicator of the activity of the sinus node is the contraction of the

atrium and the electrical phenomena connected with it it is obviously impossible to distinguish between failure to form impulses within the node and failure of a sinus impulse to reach the atrium By convention then disappearance of the P waves in the electrocardiogram through one or several cycles is called sinoatrial block

The mildest degree consists of an occasional dropping out of a P wave (Fig 74 p 77) In other instances the halving of the rate may persist for several minutes giving rise to marked bradycardia and even to Adams Stokes syndrome Persistent sinoatrial block is almost un known in children however occasional dropped beats occur relatively frequently in patients having rheumatic carditis primary myocardial disease or digitalis intoxication and also in persons without heart disease especially in the newborn

No symptoms result from this arrhythmia in children I have never seen Adams Stokes attacks resulting from it, although obviously they do occur in adults

The diagnosis usually rests on the electrocardiogram although bradycardia responding to atropine administration by a doubling of the rate should suggest it even without an electrocardiogram

Sinoatrial block does not require treatment unless the underlying heart disease needs attention

Atrioventricular Block

Atrioventricular block is an interference with the normal conduction of the impulses from the atria to the ventricles through the atrioven tricular node There are several degrees of block

First Degree Block

First degree atrioventricular block is defined electrocardiograph ically as a P R interval longer than that expected from the patient s age and heart rate It cannot be emphasized strongly enough that a pro longed P R interval does not necessarily indicate heart disease as a matter of fact, in at least one study [55] 90 per cent of the patients with first degree block had no other evidences of heart disease Among the etiologic factors should be mentioned rheumatic carditis congenital heart disease (atrial septal defect, Ebstein s anomaly) primary myo-cardial disease diphtheria digitalis and much less commonly quinidine intoxication Long P R intervals of rheumatic carditis may be reverted to normal by the administration of atropine [39] perhaps indicating the vagal rather than myocardial origin of this phenomenon First-degree heart block per se does not cause symptoms or require treatment

Second Degree Block

In second degree atrioventricular block an occasional P wave is not conducted to the ventricles at all This may happen haphazardly or it

may occur once every three four or five beats It may also be noted as a so called Wenckebach phenomenon, within a series of progressively prolonged P R intervals culminating in the complete blocking of one atrial impulse (Fig 52 p 59)

Second degree block is not common in patients without heart disease The etiologic factors responsible for first degree block are the most significant factors causing second degree block as well No treatment is necessary except as indicated by the underlying heart disease

Third Degree Block

In third degree atrioventricular block (complete heart block) the atria and the ventricles beat independently Their rates are different and in contrast to atrioventricular dissociation the ventricular rate is always the slower of the two

Physiology

The ventricular rate in children with complete heart block is considerably faster than in adults with acquired heart block The pulse rate is usually fixed between 40 and 60 beats per minute it is less than 40 in only about 10 per cent of the cases A change of more than 20 beats per minute after exercise is noted only rarely although an increase of 10 to 20 beats per minute occurs frequently If the myocardium is in good condition the cardiac output at this rate can be maintained within normal limits at the expense of an increased stroke volume Because of the increased stroke volume and the secondary peripheral vasodilatation wide pulse pressure will be noted Of twenty two patients with complete heart block studied at The Children s Hospital none had a pulse pressure of less than 30 mm of mercury while in half of the patients it was more than 50 mm

Etiology (Table 13)

Complete heart block in children is usually congenital in origin Rheumatic heart disease seldom if ever causes third degree block in this age group Of our twenty two patients two had definite cyanotic congenital heart disease (one with diminished and the other with increased pulmonary vasculature) and in two others cardiac catheterization revealed a *left to right shunt at the low atrial level (? ostium primum)* Three patients were thought on clinical grounds not to have associated heart disease (no murmurs no symptoms normal roentgenograms and electrocardiograms) In the remaining fifteen patients various associated congenital cardiac anomalies were suspected (ventricular septal defect atrial septal defect aortic stenosis and pulmonary stenosis) but in three of these right heart catheterization failed to reveal any left to right shunt

It has been assumed [6] that congenital heart block is commonly

Table 13 Etiology of Congenital Heart Block in 22 Patients

No heart disease (no murmur normal x ray and EKG)		3 patients
Definite congenital heart disease		4 patients
a Cyanotic	2 patients	
b Left to right shunt	2 patients	
Possible congenital heart disease		15 patients
No shunt at venous catheterization	3 patients	
Tentative diagnoses in others		
Atrial septal defect		
Ventricular septal defect		
Aortic stenosis		
Endocardial fibroelastosis		

associated with ventricular septal defect Much of the evidence on which this assumption is based is purely clinical namely the presence of a harsh systolic murmur at the lower sternal border From the embryologic viewpoint it seems rather unlikely that an isolated small ventricular septal defect usually located high and anterior in the membranous septum would interfere with the development of the bundle of His which runs posteriorly and is well developed before the formation of the septa [367] In fact a few cases of congenital heart block with ventricular septal defect confirmed at postmortem examination have been described in the literature but certainly far fewer than would be suspected on clinical grounds

One further etiologic factor to be mentioned is endocardial fibroelastosis which has been found at postmortem examination in patients with congenital heart block and may very well explain the clinical and physiologic findings in some of these patients

Briefly then on the basis of our own experience and a review of the literature I do not believe that ventricular septal defect is as commonly associated with complete heart block as was hitherto believed Third degree heart block may well be associated with a variety of cyanotic or acyanotic congenital heart diseases or even endocardial fibroelastosis It is also suggested from our material that complete heart block per se may not cause cardiac enlargement murmurs or electrocardiographic changes

Clinical Picture

In the majority of instances the condition is discovered during a routine physical examination although on close questioning a history of fatigue may be obtained in about half of the children In a few instances the irregularity of the heart beat or the appearance of "spells" is noted by the parent Some of these patients are underdeveloped and have a history of occasional cyanosis We have not seen true Adams Stokes attacks in any of these youngsters although one of them had convulsions possibly epileptic Two patients had evidences of congestive failure

A grade 3 or 4 harsh blowing systolic murmur was heard at the left sternal border in nineteen of our twenty two children Low frequency early or mid diastolic murmurs were also noted in more than half of these children Three patients showed no murmurs at all and had normal roentgenograms and electrocardiograms

The changing intensity of the first heart sound depending on the proximity of the QRS to the preceding P wave has been noted repeatedly and is characteristic of complete atrioventricular block An atrial or fourth heart sound often can be detected by the stethoscope and demonstrated by phonocardiograms (Fig 104 p 101)

Roentgenologic evidence of cardiac enlargement was found in more than half of our patients In most instances the enlargement was generalized involving all cardiac chambers The status of the pulmonary vasculature was variable

The electrocardiogram of course showed complete atrioventricular block in all In twelve of the twenty one patients in whom complete electrocardiograms were obtained additional abnormalities such as left ventricular hypertrophy with or without T wave changes or evidences of atrial hypertrophy were found In all instances the QRS complexes were of the normal supraventricular type Atropine or ephedrine given in an attempt to speed up the ventricular rate was followed by an appreciable rise in pulse rate in only one instance

Course and Prognosis

On the whole the course of this group of patients is rather satisfactory Only one of our twenty two patients died—a boy with tetralogy of Fallot The others lead normal active lives with few if any limitations Campbell and Thorne[76] followed up seven patients for twenty five years and observed no significant changes in cardiac status Obviously the presence and the nature of the associated cardiac anomaly determine to a great extent the ultimate fate of these children Congestive failure is more likely to be caused by the coexisting heart disease than by the heart block itself Adams Stokes attacks are common in adults

One of our two patients with cyanotic congenital heart disease has now reverted to sinus rhythm This phenomenon in patients with congenital heart disease and complete heart block has also been mentioned by Campbell and Thorne [6]

Treatment

Since most of our patients with complete heart block are relatively symptom free we did not feel justified in attempting to increase the ventricular rate In adults in whom Adams Stokes attacks are common the continuous oral administration of ephedrine may be necessary Digi

talization and other anticongestive measures may be indicated if cardiac failure occurs

CARDIAC ARREST

Complete cessation of the circulation can be regarded as the most serious arrhythmia imaginable hence it may be discussed in this chapter on the disturbances of rhythm

Close to 90 per cent of cardiac arrests are based on complete mechanical and electrical asystole in the other 10 per cent ventricular fibrillation is the underlying mechanism[3 1] Differentiation between the two is impossible without an electrocardiogram At the operating table the heart may seem to be entirely motionless on direct inspection but the electrocardiogram still may show fibrillary activity

Cardiac arrest has occurred much more frequently in children since the introduction of cardiac surgery Johnson and Kirby[178] discussing principally adults placed the incidence at about one in 5000 operations Before the era of thoracic surgery the only etiologic factor of any significance was vasovagal reflex that is sudden cardiac standstill during paracentesis or introduction of an intratracheal tube

The physiologic effects of cardiac arrest concern primarily the brain Lack of an adequate supply of oxygen to the central nervous system for three minutes or more usually results in irreversible damage (blindness convulsions severe mental deficiency) even if the circulation later is restored

The clinical recognition of cardiac arrest during operation rests on the absence of blood pressure and pulse and on the cessation of bleeding Dilatation of pupils respiratory arrest and generalized cyanosis are late signs Zoll and his associates[371] recommended the use of a cardiac monitor for instantaneous recognition of asystole during operation

The most important point in the management of cardiac arrest is its prevention Adequate preventive measures have been outlined in the section on Ventricular Fibrillation (see p 201)

If cardiac arrest does occur however early recognition and prompt treatment within three minutes according to a well rehearsed plan are of the utmost importance

The first step in the treatment of cardiac arrest is the establishment of an adequate airway through an intratracheal tube Pure oxygen (10 to 15 mm of mercury pressure) can be administered through this within less than one minute

While the airway is being introduced external stimulation of the heart may be undertaken If the chest is closed this may be accomplished (1) by pounding the left precordium (2) by the introduction of a needle through the chest wall (3) by intracardiac injection of epinephrine (0 1 to 0 2 cc of 1 1000 solution) or (4) by using a cardiac

pacemaker[371] No more than a total of two minutes should be spent in external resuscitation and establishment of an airway

In the third minute of asystole if the chest is closed—or immediately upon recognition if the chest is open—cardiac massage should be started As the massage is started procaine should be injected into the pericardium

The effectiveness of any of these procedures may best be judged by the return of a palpable pulse a measurable blood pressure and a constriction of the pupils

Once cardiac action has been started again—and only then—an electrocardiograph should be connected to the patient for accurate analysis of the underlying mechanism

If the electrocardiogram shows ventricular fibrillation defibrillatory measures should be undertaken by means of an electrical defibrillator delivering 1 5 to 2 0 ampere shocks One electrode is placed over the atria the other on the posterior surface of the heart Three shocks each strong enough to throw the entire body into a violent jerk should be delivered in succession A few seconds should then be allowed to elapse in order to judge whether normal mechanism has been re established If not massage should be resumed for a few minutes followed by repeated attempts at defibrillation In the meantime potassium chloride (5 to 10 mEq) or procaine amide (50 to 100 mg repeated every ten minutes) may also be injected into the ventricles The alternating use of defibrillation massage and drugs is recommended until normal cardiac function is resumed or for at least thirty minutes after the occurrence of the arrest

If the electrocardiogram indicates absence of any electrical activity of the heart massage should be maintained for at least sixty minutes with a good oxygen supply Administration of calcium chloride (2 to 4 cc of a 10 per cent solution) epinephrine (0 1 to 0 2 cc of a 1 1000 solution) or Isuprel (1 cc of a 1 500 solution) intracardially is also recommended

By the prompt use of these resuscitative methods a certain number of patients may be restored to life Kirby and Johnson[178] were able to resuscitate ten of nineteen adults most of whom did not have cardiac disease In larger series only 25 to 35 per cent recovery is reported Obviously a patient without heart disease responds better and more promptly than one in whom there is a cardiac defect Equally clearly the sooner the resuscitative measures are started the better the results will be

CHAPTER 10

Myocardial Diseases

Diseases affecting principally the myocardium will be discussed in this chapter. Myocardial disease thus is contrasted with rheumatic fever and subacute bacterial endocarditis (in which endocardial involvement is almost a *sine qua non*), with pericarditis and the arrhythmias (which represent purely functional derangements), and with congenital heart disease (typical examples of structural defect).

PRIMARY MYOCARDIAL DISEASE

As a purely clinical entity, primary myocardial disease is characterized by changes in the myocardium not associated with any other known disease. These patients have (1) cardiac enlargement without significant murmurs and (2) electrocardiographic abnormalities, principally changes in the S-T segments and T waves.

None of them have (1) evidences of left-to-right or right-to-left shunts, (2) pericarditis, (3) paroxysmal atrial tachycardia, (4) arterial hypertension, (5) renal disease, (6) neuromuscular diseases (Friedreich's ataxia or muscular dystrophy), or (7) infections (streptococcal infections, diphtheria, acute exanthemas, and so forth) preceding or associated with the disease. Most of these conditions can easily be excluded from the differential diagnosis, but occasionally the presence of fever, pulmonary rales, and harsh breath sounds makes the differential diagnosis between congestive failure consecutive to myocardial disease and myocardial disease secondary to pneumonitis an exceedingly difficult one. Knowing the rarity of heart failure in even the severest attacks of pneumonia, it is my belief that in most of these cases the myocardial involvement probably precedes the infection. This contention finds a practical demonstration in the beneficial effect of anticongestive measures on the pulmonary changes of these patients.

Some years ago, in order to learn the etiology of primary myocardial disease, Rosenbaum, Neuhauser, and I collected forty-five cases with clinical syndromes of the disease from the files of The Children's Medical Center.[2] Twenty-six of these children died and were examined post mortem. The five pathologic entities producing this clinical syndrome and their frequencies are listed in Table 14. It may be seen from this table that most of the patients had subendocardial sclerosis (endo-

Table 14 Autopsy Findings in 26 Cases of Primary Myocardial Disease

Disease	Number of Cases
Glycogen storage disease of the heart	3
Aberrant left coronary artery	1
Medial necrosis of the coronary arteries	2
Idiopathic myocarditis	10
Subendocardial sclerosis	10
Total	26

cardial fibroelastosis) or idiopathic (inflammatory) myocarditis (Fiedler's) [117]

Nineteen of the forty five patients are living and well What the anatomic diagnosis is in these patients can only be surmised However they are likely to fall into the category of either idiopathic myocarditis or subendocardial sclerosis because of their survival and even improvement through months and years As will be pointed out later an accurate differential diagnosis in vivo between these two conditions is practically impossible

In the following pages the five etiologic entities encountered at postmortem examination will be described first and then a discussion of our nineteen living patients will be presented Table 15 summarizes the pertinent clinical findings

Glycogen Storage Disease of the Heart

Since Pompe's original description [74] in 1933 less than forty well authenticated cases have been reported in the literature [13] The fundamental difference between cardiac glycogen disease and the hepatorenal type has been stressed by all concerned The two never overlap as a matter of fact hepatomegaly (for reasons to be given later) is probably a less common feature of glycogen storage disease of the heart than of other types of primary myocardial disease

The histologic picture is characterized by abnormally large glycogen deposits in the heart and skeletal muscles (Best's carmine stain) Few quantitative glycogen determinations on biopsy material or fresh postmortem specimens are available However all the evidence so far points to a considerable increase in the glycogen content of the heart (6 to 8 per cent of fresh weight instead of 1 per cent) and skeletal muscle (1 5 per cent instead of 0 5 per cent) [93] There is evidence that the glycogen may not be abnormal in structure since in skeletal muscle autolysis takes place at 37° C at a normal rate and in heart muscle the rate of autolysis is only slightly slower than normal It is interesting to note that the increase in glycogen content in muscle does not account for the tremendous increase in heart weight much of which is probably explained by the accumulation of water [344] There is no well accepted

Table 15 Clinical Aids in the Etiological Differentiation of Primary Myocardial Disease

Disease	Age at Onset	Family History	Congestive Failure	Remarks
Glycogen storage disease of the heart	2–6 months	Often positive	Absent	Skeletal muscle biopsy may show high glycogen content
Aberrant left coronary artery	2–6 months	Negative	Absent	Symptoms often occur in attacks chest x ray may show bulge in region of left ventricle
Medial necrosis of the coronary arteries	Less than 3 months	Negative	Absent	May have associated congenital anomalies often evidence of systemic disease especially with renal involvement
Idiopathic myocarditis	Throughout pediatric age range	Negative	Present	Gallop rhythm common congestive failure often responds well to digitalis
Subendocardial sclerosis	Throughout pediatric age range	Negative	Present	Cannot be differentiated clinically from idiopathic myocarditis

biochemical explanation for the accumulation of glycogen in the heart and skeletal muscles except that it probably involves some difficulty in phosphorylation

The disease is commonly a familial one and is usually sex linked within the same family consanguinity is frequently present The clinical picture is dominated initially by a tremendously enlarged heart with relatively few symptoms although anorexia poor weight gain cough vomiting and abscesses have also been noted Dyspnea and spells of cyanosis usually occur later Evidences of right sided heart failure are relatively rare if they occur they are terminal events At physical examination one is impressed by the extremely large heart with only slight if any murmur In the two patients whom I have had the opportunity to observe personally the mechanical compression of the lung by the tremendous heart was the most prominent physical finding The presence of macroglossia may suggest cretinism in the differential diagnosis

Results of laboratory examinations aimed at determining the adequacy of glycogen metabolism (fasting blood sugar glucose tolerance epinephrine tolerance and urinary acetone determination) are usually within normal limits in patients with glycogen storage disease of the heart The radiologic picture is characterized by diffuse cardiac enlargement normal pulmonary vasculature and a poor beat at fluoroscopy



Figure 144 Posteroanterior radiogram of a baby with glycogen storage disease of the heart *A* At birth *B* At three weeks of age

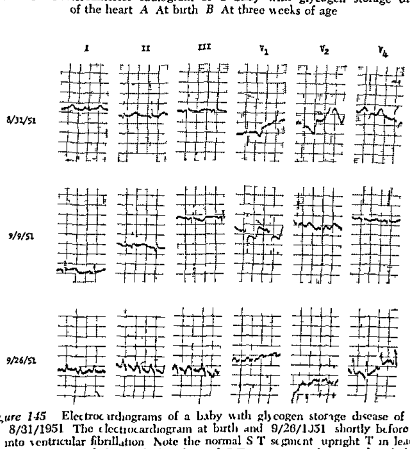


Figure 145 Electrocardiograms of a baby with glycogen storage disease of the heart 8/31/1951 The electrocardiogram at birth and 9/26/1951 shortly before he went into ventricular fibrillation Note the normal S T segment upright T in lead I in the first tracing and the markedly elevated S T segment in the second and third tracings in lead I Also observe the pointed T in the last tracing with normal T waves in the first and second ones Finally note the upright T in lead V_4 in the tracing of 8/31/1951 with inverted T wave on 9/9/1951 and 9/26/1951

Electrocardiographically evidences of left ventricular hypertrophy with changes in the S T segments and T waves are noted

A personal observation regarding the time of development of these changes is of particular interest At The Children's Hospital we had cared for two siblings with glycogen storage disease The family notified us immediately after delivery about the arrival of a third child Physical and radiologic examinations and electrocardiographic studies showed the newborn to be perfectly normal in all respects Cardiomegaly and electrocardiographic changes first appeared when the child was three weeks old and death in ventricular fibrillation ensued at four weeks (Figs 144 and 145)

According to the literature symptoms never occur at birth but are always present by the sixth month of life and sudden death without congestive failure almost always occurs before the child has reached one year of age Therapy is usually of no avail

Aberrant Origin of the Coronary Artery

This is another rare anomaly less than fifty cases have been reported Because of a developmental anomaly in the embryologic division of the common arterial trunk the left coronary artery originates from the pulmonary artery rather than from the aorta In other and even rarer cases the right or even both coronary arteries may originate from the pulmonary trunk

Physiologically the handicap is twofold the blood perfusing the heart has the low oxygen content of the pulmonary artery rather than the full saturation of the aortic blood and the perfusion pressure after the neonatal period is only about a quarter of that of the systemic circuit That the first handicap may not be the more important one is suggested by the observation that patients with cyanotic congenital heart disease whose coronary artery blood sometimes has an extremely low oxygen content may do very well for many years The crucial role of the low perfusion pressure of the coronary artery is suggested by the fact that infants rarely get into severe difficulties with this condition until they are some months old this may be explained by the fact that the pulmonary arterial pressure is relatively high in young infants and undergoes a general decrease as the baby gets older

The clinical picture was first described by Bland White and Garland[38] Symptoms usually occur between the second and sixth months of life and death ensues before one year of age The characteristic symptoms are episodes of colicky pain pallor cyanosis tachycardia tachypnea and perspiration These are likely to occur at feeding time and last for a few minutes they resemble anginal attacks of adults Between these spells the infant is usually well although the heart is large An enlarged liver indicating right sided congestive failure is rarely and only terminally noted

Radiologically generalized cardiac enlargement characteristic of the entire myocardial disease group may be seen The only distinguishing *feature on the* radiogram may be the occurrence of a localized aneurysmal bulging in the area of the left ventricle

The electrocardiograms are characterized by deep inversion of the T waves in the standard limb leads and left precordial leads These patterns much like the x ray picture are no more characteristic of this condition than of the other members of the primary myocardial disease *group* If however in addition to the T wave changes deep Q waves occur in lead I and QS complexes in the left chest leads indicating a necrosis or a hole in the myocardium then the diagnosis of anomalous *origin* of the coronary artery seems justifiable on the basis of the electro cardiographic picture alone

No effective treatment has been devised so far although trans plantation of the coronary artery has been suggested[1, 8]

Medial Necrosis of the Coronary Arteries

This is the rarest of the five conditions being discussed less than thirty such cases have been reported Pathologically a generalized *vascular* disease with medial necrosis intimal proliferation and eventu ally arterial occlusion is the underlying abnormality In addition to the coronary arteries the pulmonary and the renal vessels are also likely to be involved

As may be expected this is an extremely severe condition usually leading to death at less than three months of age The symptoms are anorexia vomiting respiratory infection pallor cyanosis and terminally congestive failure The roentgenograms and electrocardiograms are much the same as have been described in the previous two conditions but the deep Q waves characteristic of anomalous coronary arteries are usually absent

The patients in this group are distinguished from those having other types of primary myocardial disease by the presence of associated con genital anomalies and by the involvement of the renal arteries

Idiopathic Myocarditis

In contrast to the type of myocarditis which occurs with bacterial and virus diseases[30 335] no definite etiologic agents have been implicated in this group of patients It is my conviction however that with the improvement of diagnostic techniques it will be possible to implicate definite pathogenic agents in many of these cases Strong support of this view may be found in the "epidemic occurrence of myocarditis in 140 newborns in Munich described by Stoeber[3 3] in 1952

This is not nearly as rare a condition as the three discussed previ ously More than 100 cases have been reported in the literature since

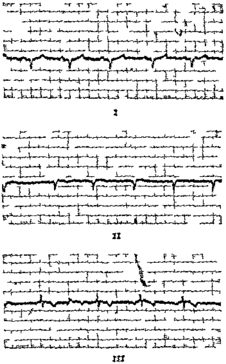


Figure 146 Electrocardiogram of patient with idiopathic myocarditis Note the low voltage and the slurring of QRS complexes in leads I to III Also observe flat T_2 and inverted T_3

Fiedler's[117] original paper in addition to the 140 described in the Munich epidemic

The pathologic picture depending on whether one is dealing with an acute subacute or chronic phase of the disease may show *interstitial* edema diffuse perivascular round cell infiltration or even fibroplastic granulomatous changes

The clinical picture is usually characterized by the sudden onset of left and right sided congestive failure A child usually six months old or older who was perfectly well a few hours before becomes desperately ill with dyspnea pallor cyanosis and a hacking cough The temperature is moderately elevated Pulmonary rales appear and the *liver is enlarged Definite cardiac enlargement is present and the heart* sounds are muffled Gallop rhythm without a murmur is noted Characteristically the symptoms and signs are not episodic the child is continually and severely ill

Roentgenograms show features common to the entire primary myocardial disease group The electrocardiographic changes are also characteristic of the entire group low voltages and conduction disturbances

(atrioventricular and intraventricular) may be present more commonly in this condition than in the others (Fig 146) Conversely severe left ventricular hypertrophy may be somewhat rarer

In the differential diagnosis pneumonitis septicemia and paroxysmal tachycardia should be considered In some cases treatment may be extremely effective Although conclusive proof is missing I believe that many if not most of our successfully treated cases of primary myocardial disease fall into this category Oxygen and antibiotic therapy should be given routinely The principal drug to be used however is digitalis administered in average or slightly lower than average doses and maintained for several months or even years (Table 12 p 184) The response is just short of miraculous in many instances Careful electrocardiographic control is necessary because some of these children are susceptible to digitoxicity

Within recent years steroids have been used in some patients with the presumptive diagnosis of idiopathic myocarditis I have observed two children treated with cortisone who made good recoveries At present I see no reason to abandon the therapeutic regimen consisting of digitalis with supportive treatment However as more is learned about hormone therapy it may well be used with increasing frequency

Subendocardial Sclerosis (Endocardial Fibroelastosis)

This is one of the commonest types of primary myocardial disease Dennis and his associates[9] in 1953 collected 149 cases from the literature

Pathologically the condition is characterized by a milky appearance of the parietal endocardium involving principally the left atrium and the left ventricle Frequently there is associated involvement of the mitral or aortic valve or both Involvement of the right heart chambers is rare

Histologically the endocardium is two to three times thicker than normal with endocardial and subendocardial fibroelastic proliferation a picture similar to that seen in "collagen disease" The myocardial fibers are often hypertrophied but signs of inflammation are strikingly absent

Detailed physiologic studies are lacking Using catheterization of the right side of the heart we studied three patients who had a clinical diagnosis of fibroelastosis All of these had mild elevation of "pulmonary capillary" and pulmonary arterial pressures Similar observations have been made by others[3] If the observations made on patients with myocardial fibrosis and constrictive pericarditis by Burwell and his associates[67] can be considered to apply to this group it may be assumed that the pathologic process results in an elevation of the diastolic pressure and impairment of systolic ejection of the left ventricle with a narrowing pulse pressure It results in "constrictive endocarditis" similar physio

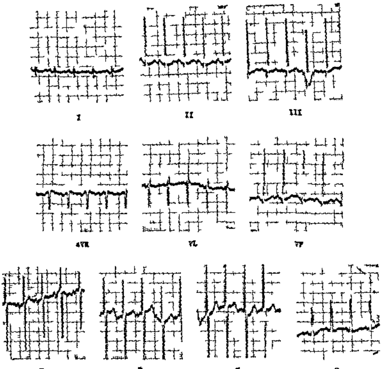


Figure 147 Electrocardiogram of a patient with endocardial fibroelastosis Note the evidence of left ventricular hypertrophy in tall R_2 and R_3 and R aV_F Also notice the low T waves in V_6.

logically to constrictive pericarditis The poor diastolic emptying of the left ventricle in a case of endocardial fibroelastosis has been demonstrated angiocardiographically by Gasul and his associates[1 8]

The etiology of subendocardial sclerosis is not known Farber and Hubbard[114] in 1933 emphasized the inflammatory nature of the condition Within recent years it has been regarded more and more as a developmental anomaly because of its occasional familial occurrence (we have seen it in two sets of siblings) and its frequent association with other types of congenital heart disease[43 105 148] The question must be raised whether the cases in adults reported in the literature[330] are basically different from those seen in infants and newborns I believe that the cases in adults in contrast to the primary endocardial fibroelastosis of infants may well be secondary to coronary disease myocarditis or metabolic disturbances Convincing proof for this hypothesis is missing

Often the clinical picture of subendocardial sclerosis cannot be distinguished from that of other members of the primary myocardial disease group the similarity toward primary myocarditis is particularly striking Approximately 50 per cent of these patients have symptoms

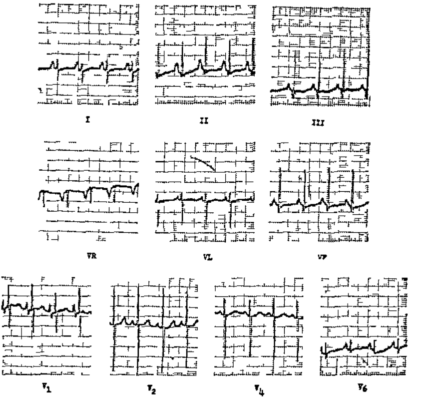


Figure 148 Electrocardiogram in patient with endocardial fibroelastosis Note the marked atrial hypertrophy as revealed in lead II aV$_R$ and V$_1$

within the first six months of life and according to Blumberg and Lyon[43] in 95 per cent of the cases symptoms become manifest within the first year However *in some cases of pathologically confirmed sub endocardial sclerosis the patients were completely asymptomatic until they were five to six years of age* Left and right sided congestive failure dominates the picture The children may be cyanotic but the cyanosis may improve considerably when 100 per cent oxygen is administered Pulmonary rales and dullness with a hacking cough suggest the pres ence of pneumonitis or pulmonary edema Peripheral edema is rare but hepatomegaly and distended neck veins are common On auscultation the heart sounds are clear gallop rhythm is not too common and mur murs are faint or do not exist The infants are usually underdeveloped and undernourished

Electrocardiographically evidences of left ventricular hypertrophy with T wave changes can be seen (Fig 147) Marked right atrial hy pertrophy may be present (Fig 148) Low voltage (Fig 149) and conduction disturbances are rare

Radiologically there is an enlarged heart with involvement of all

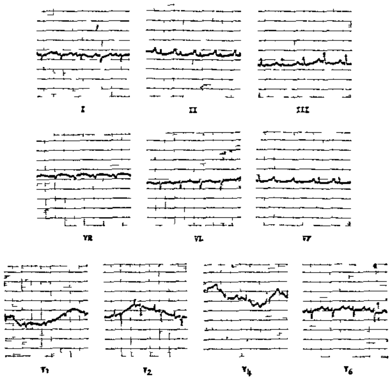


Figure 149 Low voltage tracing in a patient with endocardial fibroelastosis

chambers and a poor beat under the fluoroscopic screen There are usually marked evidences of pulmonary congestion or infection (Fig 150)

Treatment should consist of anticongestive measures principally the administration of digitalis and mercurials with oxygen and antibiotics The similarity of the histologic findings to those seen in collagen disease has suggested the use of steroids in this condition however I am not acquainted with any sizable series of patients treated with hormones

Results of treatment are extremely hard to judge Some patients with a clinical picture practically identical to the one described respond well to digitalization others with the same clinical profile die in spite of vigorous treatment So far there are no reliable criteria to indicate which patients will or will not respond to present day therapeutic regimens However it seems that the younger the infant and the more severe the respiratory symptoms the worse the prognosis is likely to be Also if after an initially favorable response to treatment, the symptoms recur the ultimate prognosis seems worse Patients who eventually get better usually respond promptly and have no recurrences



Figure 150 Posteroanterior and oblique radiograms in a patient with subendocardial sclerosis

Patients with Primary Myocardial Disease Who Recover

As indicated at the beginning of this section on primary myocardial disease nineteen of the forty five patients with primary myocardial disease seen at The Children s Hospital are still living and well [9] Since no anatomic basis for a definite diagnosis in these patients exists they cannot be discussed under any of the previous subheadings At the same time on the basis of the clinical definition of the syndrome they do belong in this general category They all had cardiac enlargement without significant murmurs and changes in the S T segments and T waves in the electrocardiogram

If one were to speculate about *the anatomic diagnosis in these* patients one would feel compelled to exclude glycogen storage disease anomalous coronary arteries and medial necrosis of the coronary arteries by virtue *of the fact that these children* recovered There is practically

no instance in the literature of a patient surviving beyond the first year of life when it was proved that he had any one of these three entities Therefore these living patients must belong to the idiopathic myocarditis or subendocardial sclerosis group unless they represent another entity outside the scope of the etiologies suggested by our pathologic material

Patients with reversible" primary myocardial disease are encountered at our hospital about as frequently as those with paroxysmal atrial tachycardia—that is two or three times a year The age at which symptoms begin varies from birth to seven years Almost half of the patients were one year old or older when they first became sick This is the most suggestive evidence that many of these youngsters belong in the idiopathic myocarditis group

The clinical picture in the typical case is characterized by the sudden appearance of symptoms of congestive failure—dyspnea cyanosis anorexia and vomiting The picture may resemble that of pneumonia especially if cough and fever are also present In rare cases the onset is insidious congestive failure being preceded for several months by anorexia failure to gain weight irritability and profuse perspiration

On examination many of these children are well developed and well nourished some are actually chubby this again suggests that the etiology is less likely to be a congenital anomaly than an acquired inflammatory disease Generally these patients look very ill their color is pale with cyanosis their respirations are rapid or labored and peripheral edema may be present Rales are commonly heard and there is almost always significant hepatomegaly The heart sounds are distant and "mushy with a gallop rhythm There are no significant murmurs deformity of the left side of the chest may be present but is not too common The temperature may be slightly elevated but a fever higher than 102 F is rare The roentgenograms and electrocardiograms cannot be distinguished from those in the rest of the primary myocardial disease group

Response to treatment is dramatic in these patients Thirteen of our nineteen children received digitalis and one received cortisone In addition Mercuhydrin was administered to the children with peripheral edema All received general supportive measures such as oxygen and antibiotics Within twenty four hours considerable clinical improvement occurred and in one week these youngsters were practically asymptomatic The electrocardiograms returned nearly to normal within six to twelve months (Figs 151 152) the hearts were considerably reduced in size within six months but did not return to normal for one or two years Digitalis therapy was routinely maintained for six months after which an attempt usually was made to suddenly discontinue the drug If the child started having symptoms again the administration of the glycoside was resumed and continued for another year at least

It should be mentioned here that not only are these patients bene

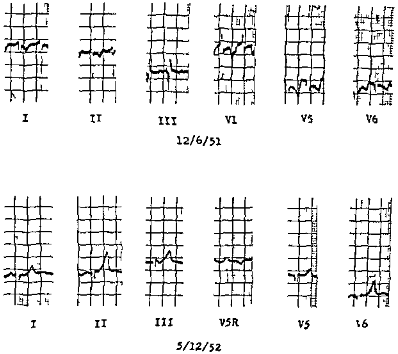


Figure 151 Electrocardiogram of patient with recovered primary myocardial disease The upper tracing shows S T and T wave changes in T_1 V_5 and V_6 whereas the complexes are entirely normal on the lower tracing

fited greatly by digitalization but they also show toxic effects readily Electrocardiographic evidences of digitoxicity are often seen with average or lower than average doses of the glycoside this is most emphatically not an absolute indication for discontinuance of the drug if good therapeutic effects are accomplished Rather an attempt should be made to diminish the dose to a point at which maximal therapeutic effects are achieved with minimal effects of toxicity

Although the short term prognosis for a child who shows dramatic initial improvement to anticongestive measures is good obviously the long term outlook cannot be predicted Considerable encouragement may be gained from the fact that many of these children who have been followed up for five years or more are now asymptomatic and have normal hearts as judged by physical examination roentgenograms and electrocardiograms These children are being encouraged to lead perfectly normal lives although I am inclined to discourage participation in competitive athletics in high school or college

A personal and wholly unsubstantiated view in regard to the etiology of the disease in these children is that idiopathic myocarditis rather than subendocardial sclerosis is the usual pathologic change I can scarcely believe that the thick layer of constrictive subendocardial elastic tissue can disappear as a result of anticongestive measures any

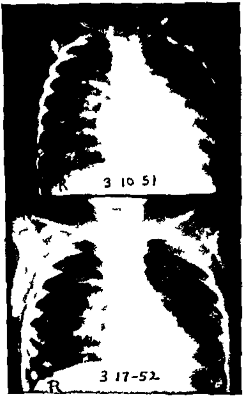


Figure 152 Upper the heart in a patient at the height of his primary myocardial disease Bottom one year later after complete disappearance of signs and symptoms of heart disease

more than constrictive pericarditis is "cured" by such means The age and good nutritional state in these patients also suggest idiopathic myo carditis rather than subendocardial sclerosis

MYOCARDIAL DISEASE ASSOCIATED WITH DIPHTHERIA

The heart is affected in approximately 10 to 20 per cent of all pa tients who have diphtheria However about 50 per cent of those who succumb to diphtheria have associated myocardial involvement[3 3] *Diphtheria bacilli are not found in the heart and apparently the cardiac* effects of the disease are caused by exotoxins liberated by the bacillus The typical pathologic picture is one of fatty and hyaline degeneration with only a small degree of cellular infiltration

Circulatory derangements associated with diphtheria may present themselves either in the form of early peripheral collapse or late myo cardial injury

Circulatory collapse is probably caused by the effects of the toxin on the peripheral vasculature and is characterized by evidences of shock

The patient's blood pressure drops precipitously, the pulse rate rises and the skin looks gray and feels cold and clammy This state of shock usually occurs toward the end of the first week of the disease the earlier it occurs the more severe is the prognosis There are usually no electrocardiographic changes Treatment consists in serum therapy of the underlying disease and shock treatment of the vasomotor collapse by administration of plasma Neosynephrine or Levophed

Diphtheritic involvement of the myocardium occurs most commonly toward the end of the second or the beginning of the third week usually when convalescence is well under way One of the few personal recollections I have of this condition is that of a boy six to seven years old who was convalescing from diphtheria and playing happily in his crib at the hospital when he suddenly became pale cried out and died He may have had an Adams Stokes attack or he may have succumbed to ventricular fibrillation Fortunately things are not always this sudden or catastrophic Electrocardiographic changes—specifically depression of the S T segment T wave inversions atrioventricular block and bundle branch block—all may be seen at the onset of the myocardial involvement The conduction disturbances are rare but relatively specific to diphtheria and they signify a more ominous prognosis Changes in the S T segments and T waves on the other hand may be seen in practically any kind of myocarditis with a less severe general outlook

At physical examination the heart may show either severe tachycardia or bradycardia There are no murmurs The heart sounds are distant and gallop rhythm may be present In rare instances right sided congestive failure occurs terminally

The prognosis of diphtheritic myocarditis is grave the mortality is placed at about 50 per cent by most authors [71] If the patient survives recovery is usually complete Levine[68] believes that many of the cases of complete heart block first manifesting themselves when the patient is beyond the age of fifty are late sequelae of diphtheritic myocarditis White[340] denies this contention

Treatment is apparently not satisfactory Serum if not administered before can be given at this stage but the dangers of serum reaction must be considered carefully indeed this may prove to be catastrophic in a patient with a weakened and irritable myocardium Digitalis is not recommended except in the rare instances of frank congestive failure when cautious administration of a rapidly excreted preparation may be advisable if one is fully cognizant of the dangers involved in its use

MYOCARDIAL DISEASE ASSOCIATED WITH FRIEDREICH'S ATAXIA

Although the chance of a pediatrician in his general practice ever encountering a case of Friedreich's ataxia is remote those of us working in a children's hospital with an active neurologic service see it fairly frequently Doctor Bronson Crothers formerly head of the Neurological

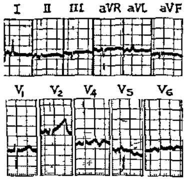


Figure 153 Electrocardiogram in patient with Friedreich's ataxia Inverted T in lead I aV V_5 and V_6 with long Q T interval

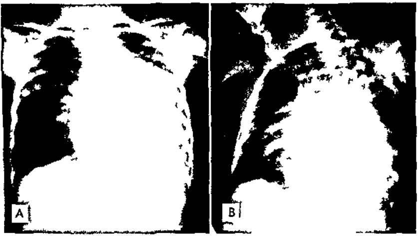


Figure 154 Posteroanterior and left anterior oblique radiograms in patient with Friedreich's ataxia and pericardial effusion

Service of The Childrens Hospital in Boston first drew my attention to this entity

Friedreich's ataxia is a hereditary familial disorder characterized by cerebellar and posterior column ataxia weakness areflexia and terminal dementia Characteristic skeletal deformities include marked scoliosis and high arched feet The disease begins during childhood and death usually occurs at puberty or in the early twenties The age at onset as well as the age at death may be characteristic for one family

Friedreich's original series in 1863 included six patients five of whom had clinical evidences of heart disease Since then a number of authors have drawn attention to this association[111 4 9] Apparently electrocardiographic abnormalities—particularly left ventricular S T and T wave changes (Fig 153)—are the most common evidences of cardiac involvement occurring in at least one third of all cases of Friedreich's

ataxia Arrhythmias—parasystole and atrial fibrillation—have been described Pericardial effusion has been seen (Fig 154) and anginal pain occurs Congestive failure is rare

The few pathologic studies available indicate the prevalence of coronary arterial changes and evidences of myocardial fibrosis

No treatment is available for either the cardiac or the neurologic aspects of this disease The electrocardiogram may be a useful diagnostic tool however Whether these patients die because of the cardiac involvement or not is difficult to tell The terminal stages of the disease are characterized by a virtual disintegration of the organism The few postmortem observations described in the literature do suggest however that myocardial fibrosis with or without coronary disease may at least be a significant contributing factor in the death of these patients [54, 99]

MYOCARDIAL DISEASE ASSOCIATED WITH PROGRESSIVE MUSCULAR DYSTROPHY

Another heredofamilial disease causing wasting and even paralysis of the skeletal muscle is progressive muscular dystrophy This is a rare disease and cardiac complications have been reported with various frequencies by various authors I am aware of only one case within the past six years at The Children s Medical Center a six year old boy with progressive muscular dystrophy had definite evidences of heart disease (Figs 155 156) He was known to have had the disease since he was two years old and he died in congestive failure Postmortem examination

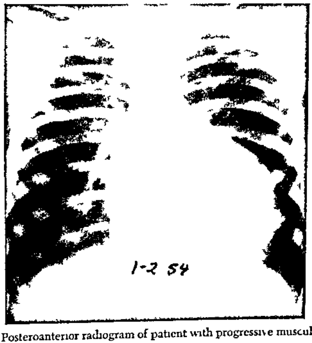


Figure 155 Posteroanterior radiogram of patient with progressive muscular dystrophy

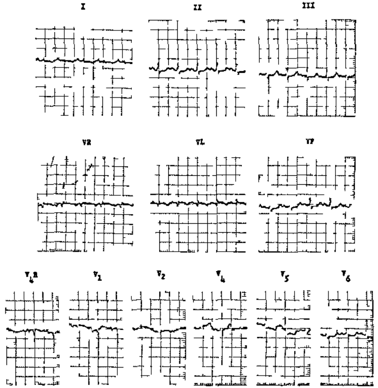


Figure 156 Electrocardiogram of patient in Figure 155 Note the low voltage standard limb leads inverted T in lead I depressed S T in aV_F and depressed S T in V_5 and V_6 with diphasic T in V_5 and V_6

revealed marked cardiac enlargement with endocardial thickening of the left ventricle and the left atrium Microscopic study showed fatty degeneration scarring and lymphocytic infiltration

Two interesting papers have been published recently on this subject [96, 330] The consensus seems to be that about 50 per cent of the cases examined post mortem show some degree of involvement of the myocardium similar to that seen in the striated muscle Among the clinical manifestations of dystrophic heart disease congestive failure as well as tachycardia arrhythmias T wave changes and abnormal Q waves should be mentioned A temporary remission of the congestive failure can be achieved in these patients with anticongestive measures

MYOCARDIAL DISEASE ASSOCIATED WITH INFECTIONS

There are two interesting and diametrically opposing trends in pediatrics today in relation to the significance of cardiovascular involvement in acute infections

In the older textbooks "toxic myocarditis" was blamed for the death of babies with bronchiolitis bronchopneumonia and acute infectious

diseases including the exanthemas There was talk about scarlet fever heart and prophylactic digitalization was widely recommended Within the past twenty five years most pediatricians have become rather skeptical about the validity of these observations on toxic myocarditis" It may well be that the antibiotics are successfully combating infections to the point at which myocardial involvement no longer occurs It is also possible that the widespread use of accurate diagnostic tools has made us interpret our observations more correctly Whatever the reason most pediatricians today are able to treat infectious diseases successfully without the use of digitalis or diuretics and by and large people have stopped worrying about toxic myocarditis It has also been learned that if the circulatory system is involved in acute infections the result is usually peripheral vascular collapse a condition requiring shock treatment rather than anticongestive measures

The opposing trend was started by the pathologists especially Saphir and his associates[140] who found myocardial changes at post mortem examination in a large number of persons who had died from infectious disease They and others brought forward histologic proof of myocardial involvement in tuberculosis hepatitis poliomyelitis encephalitis pneumonia tularemia meningococcemia infectious mononucleosis rickettsial diseases influenza mumps yellow fever and everything else one can think of These pathologic studies were followed by widespread electrocardiographic studies in patients with infectious diseases They showed transient T wave changes and some intraventricular conduction disturbances in a large percentage of children who had poliomyelitis measles hepatitis and infectious mononucleosis

Although the presence of histologic or electrocardiographic changes in infectious disease can scarcely be doubted their interpretation requires considerable caution It should be remembered that most of these patients have a high fever their nutrition may be poor and the peripheral circulation is certainly affected In poliomyelitis anoxia and difficulties in the autonomic nervous system play a significant role Finally with all this laboratory evidence the clinical proof of insufficiency of the circulation in terms of either congestive failure or significant arrhythmia is lacking in the vast majority of instances

For practical purposes it is well to treat acute infections without too much concern for the myocardium unless definite evidences of significant arrhythmia or congestive failure appear On the basis of the evidence available in the literature it is doubtful whether the state of the myocardium influences the prognosis of infectious diseases today in the vast majority of instances One exception to this statement of course is subacute bacterial endocarditis and possibly also infectious mononucleosis The latter a protean disease may produce serious cardiac damage similar to that caused by rheumatic carditis [13]

MYOCARDIAL DISEASE ASSOCIATED WITH ACUTE GLOMERULONEPHRITIS

Cardiac involvement in acute glomerulonephritis has been of considerable interest during the past two decades Pediatricians who see children with acute glomerulonephritis are well aware of the possibility of the sudden catastrophic onset of pulmonary edema which terminates fatally within a few hours

It is clear that although the majority of patients with acute nephritis have arterial hypertension this alone cannot be blamed for cardiac failure since the relationship between the two is not close Elevated venous pressure and increased blood volume have been found in a number of instances and these indubitably contribute significantly to the circulatory embarrassment Gore and Saphir described significant myocardial changes at postmortem examination in 10 per cent of their series of patients who died after acute or subacute glomerulonephritis[141]

How often cardiac failure actually occurs in glomerulonephritis is a matter of considerable controversy Ash and her associates[14] described

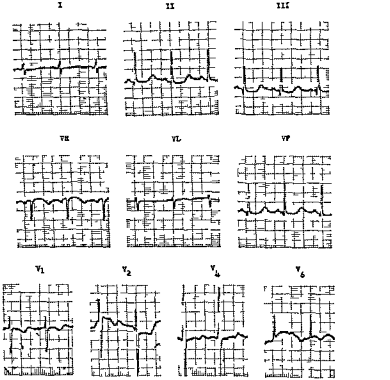


Figure 157 Electrocardiogram in patient with severe acute glomerulonephritis Note the S T segment changes in leads II III aV_F and V The T waves are inverted throughout the chest leads

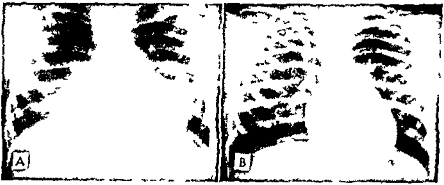


Figure 158 Posteroanterior radiograms of a patient with acute glomerulonephritis The two films were taken three weeks apart and the second one shows marked diminution of the heart size and of the pulmonary congestion

evidences of congestive failure in twenty two of fifty patients they studied while Peters[2 0] stated that congestive failure is common and that the edema of acute nephritis is due principally to this mechanism I believe that nephritic edema is probably due to two factors the increased venous pressure secondary to cardiac insufficiency and the low plasma proteins inherent in renal disease

Electrocardiographic changes (Fig 157) especially depression of the S T segment and inversion of the T wave are seen in almost 75 per cent of the patients who have acute glomerulonephritis[14] Prolonged Q Tc is seen occasionally

Radiologic evidences of cardiac failure are cardiomegaly and pulmonary congestion which rapidly clears as the patient improves (Fig 158)

The treatment of cardiac failure in acute glomerulonephritis should be aimed at reducing the load on the heart and improving myocardial efficiency The former can be achieved by eliminating the hypertension with *magnesium sulfate or Apresoline (or even Protoveratrine in an emergency)* and by trying to reduce the circulating blood volume with a low salt diet Venesection and application of a tourniquet to the extremities may even be necessary if pulmonary edema develops *Digitalis in a rapidly excreted form may be used to improve the myocardial* effectiveness but mercurial diuretics should be avoided for obvious reasons

The prognosis of acute glomerulonephritis is good in 95 per cent of the cases The circulation is restored to normal after the disease has subsided

CHAPTER 11

Diseases of the Pericardium

Classification

Pericardial disease is usually a manifestation of some systemic condition except for rare traumatic pericarditis or the presence of an isolated pericardial cyst it never occurs as an isolated disease

For practical purposes unless proved otherwise pericardial involvement indicates rheumatic carditis in children Other conditions involving the pericardium are in a sequence indicating decreasing frequencies (1) uremia (2) pneumonia with left sided empyema (3) viral disease probably caused by coxsackie virus (4) rheumatoid arthritis (5) tuberculosis (6) neoplastic metastasis (7) primary pericardial tumor or cyst and (8) constrictive pericarditis As a ninth and final point one could mention postoperative pericarditis the frequency *of which depends obviously on the number of cardiac explorations* performed in a particular institution

Pathology

Pathologically speaking pericardial disease can be divided into acute and chronic forms Acute pericarditis according to the nature of the exudate may be serous fibrinous serofibrinous purulent hemorrhagic or neoplastic Chronic pericarditis always involves fibroplastic proliferation The various clinical entities differ in the extent and nature of the involvement

Physiology

The physiologic effects of pericarditis depend to a great extent on the competence of the underlying myocardium and the nature volume and speed of accumulation of the exudate If the myocardium is intact and the serous exudate accumulates slowly several hundred cubic centimeters of intrapericardial fluid may be tolerated without obvious interference with the circulation On the other hand in rheumatic pancarditis the sudden accumulation of only 100 to 200 cc of exudate may cause serious circulatory embarrassment

Cardiac tamponade is a clinical syndrome caused by the accumula

Table 16

PHYSIOLOGIC EFFECTS & COMPENSATORY MECHANISMS IN CARDIAC TAMPONADE

IMPAIRED VENTRICULAR FILLING	⟶	INCREASED PRESSURE IN RA & SYSTEMIC VEINS
↓		
DIMINISHED STROKE VOLUME	⟶	SINUS TACHYCARDIA
↓		
DROP IN SYSTEMIC SYSTOLIC PRESSURE	⟶	INCREASE IN SYSTEMIC RESISTANCE
↓		
NARROW PULSE PRESSURE		

tion of fluid within the pericardial sac Obviously the more rapidly this fluid accumulates the larger the amount and the more rigid the pericardium the more serious the consequences will be A number of direct effects of the compression of the heart and a number of compensatory mechanisms are involved in the causation of the clinical syndrome (Table 16) Perhaps the first thing that happens is that the ventricles cannot fill adequately during diastole because of the external pressure of the fluid To counteract this the right atrial and systemic venous pressures increase Since this compensatory mechanism is not completely successful and the stroke volume is still relatively small, an attempt to maintain an adequate cardiac output is made by increasing the heart rate Finally to obviate the drop in systolic blood pressure caused by the inadequate cardiac output the diastolic pressure is raised by peripheral vasoconstriction thus achieving a satisfactory mean pressure at the expense of a narrow pulse pressure It may be worth emphasizing that the symptoms most commonly identified with cardiac tamponade such as increased venous pressure increased pulse rate and narrow pulse pressure all represent compensating mechanisms and therefore should not be "corrected" lest the cardiac output drop even further For this reason the treatment of cardiac tamponade consists solely in evacuation of the fluid Therapeutic maneuvers aimed at lowering the venous pressure (venesection) slowing the pulse rate (digitalis) or decreasing the peripheral resistance (external heat) not only are ineffective but occasionally may even prove harmful

CLINICAL PICTURE

On the whole acute dry or fibrinous pericarditis of the "bread and butter" type may be uncomfortable and painful indeed but it seldom causes failure of the circulation Conversely accumulation of a serous exudate of 200 cc or more especially if it develops rapidly obstructs the work of the heart noticeably and causes cardiac tamponade Accumulation of purulent pericardial exudate may cause not only tamponade, but also empyema like symptoms characteristic of a septic focus

Pericardial pain if present, may be of two kinds One is the dull aching nauseous poorly localized "visceral variety involving the pre

cordium and probably resulting from the stretching of the pericardial sac The other is a sharp stabbing referred type of pain localized to the left chest shoulder neck and scapula it is exaggerated by lying down and relieved by sitting up This type of pain is probably the result of the involvement of the diaphragm and the pleura since there are no sensory nerves in the pericardium proper[77]

A patient with acute rheumatic fever who suddenly starts complaining of precordial or referred pain is almost certain to show evidences of pericarditis within a few hours or days Shortness of breath grunting respirations bothersome dry cough and orthopnea with a tendency to lean forward are all characteristic symptoms of the patient with pericardial disease The symptoms of pericardial tamponade are those of acute congestive failure in a patient with pericarditis Large effusions may cause swallowing difficulties The symptoms of septicemia are characteristic of purulent pericarditis

On physical examination the most typical feature of pericardial involvement is the presence of a friction rub The characteristics of this have been mentioned previously *it is not restricted to any heart cycle or any point of auscultation and it may be evanescent in character* It should be pointed out that it may on occasion be difficult to differentiate a friction rub from a high frequency early diastolic blow the rub may be heard best with the patient sitting up or leaning forward and with the bell chest piece pressed hard against the precordium Occasionally clear cut synchronism between respiration and the friction rub is noted in which case probably a pleuropericardial friction rub is present Although friction rub is best heard in patients with dry fibrinous pericarditis its presence does not exclude the existence of a large pericardial effusion If the pericarditis is of rheumatic origin the heart sounds are rather muffled and significant murmurs are always present

An increase in cardiac dullness is usually demonstrable and the rather overrated Ewart s sign (a triangular dullness below the angle of the left scapula) is continuously sought for by medical students to the great delight of their instructors A possibly more useful sign is the fact that in spite of the large size of the heart the cardiac impulse is quiet and hypodynamic

Cardiac tamponade—an obstructive amount of fluid within the pericardial sac—manifests itself in distention of the veins in the neck enlargement of the liver and peripheral edema The systolic blood pressure is lower than normal and the pulse pressure is narrow One of the most useful signs of cardiac tamponade is the paradoxical pulse consisting in a drop in systolic pressure of more than 10 mm of mercury at the end of deep inspiration Frequently this may be noted by simply feeling the radial pulse but accurate evaluation is possible only with the sphygmomanometer These patients generally appear anxious and distressed they are cyanotic and dyspneic and look terribly sick

Table 16

PHYSIOLOGIC EFFECTS & COMPENSAT
IN CARDIAC TAMPO

IMPAIRED VENTRICULAR FILLING →
↓
DIMINISHED STROKE VOLUME —
↓
DROP IN SYSTEMIC SYSTOLIC PRESSURE —
↓
NARROW PULSE PRESSURE

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hortening and widening of
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cteristically the cardiac enlargement
d disappears after treatment or even
At cardiac fluoroscopy the pulsations
changing the patient s position from
of a tilt table) the size and shape of
well this test has been described in
of no particular assistance to us By
of contrast material (angiography) into
ventricle the presence of an extracardiac
identified This is the only way by which
erentiated from massive cardiac enlarge

changes of pericarditis have already been
30) Briefly summarized they are (1) low
the S T segment without reciprocal relation
and (3) T wave inversion which appears last
ditis and may persist for several months after
ricardial involvement have disappeared
y way to find out whether pericardial effusion
the pericardial sac by either paracentesis or
Children s Medical Center the Surgical Service
racotomy with a small incision is on the whole
n blind exploration of the pericardial sac with a
llent team of surgeons anesthetists and operating
continuous attendance this type of arrangement has
h the past years At the same time at another hos
large personnel trained in cardiovascular surgery the
pericardium by needle may be an extremely useful
edure The site of pericardial exploration should be at
side the left border of the cardiac dullness at the fifth
or occasionally in cases of large and presumably
on it should be at the angle between the xiphoid and
costal margin

gical asepsis should be observed The patient should
hine subcutaneously fifteen minutes before the start of the
Local anesthesia with 2 per cent procaine is used A 3 inch
ocar (number 16 to 18) connected with a 20 cc syringe

is used for a diagnostic tap If the approach is through the fifth inter costal space the needle should be directed to the right and toward the spine whereas in the xiphoid approach one should proceed upward and backward to the left The needle enters between the ribs and it is usually possible to feel it suddenly penetrate the pericardial sac 2 to 4 cm from the skin Usually only 20 to 40 cc of fluid are removed for diagnostic purposes If tamponade is present the removal of 150 cc or more may be a lifesaving maneuver In this case the use of a three way stopcock may prevent air from penetrating the pericardial sac this saves the trouble of detaching and attaching the needle continuously

Differential Diagnosis

The differential diagnosis of pericardial disease is not a difficult one In children the only great problem is in distinguishing pericardial ef fusion from a large heart associated with one of the myocardial diseases Features common to both are (1) congestive failure (2) large feebly pulsating heart without murmur and (3) changes in the S T segments

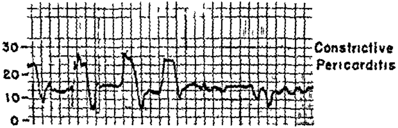


Figure 159 Right ventricular pressure curves of patient with constrictive pericarditis

and T waves on the electrocardiogram The presence of pulsus paradoxus is the deciding factor in favor of pericardial effusion If pulsus paradoxus is not present intensive decongestive measures should be attempted for one to two days keeping a careful record of the pulse pressure blood pressure and heart size If dramatic improvement does not occur within a short time or if the heart enlarges rapidly with narrowing pulse pres sure and diminishing systolic pressure then either the introduction of a cardiac catheter or the exploration of the pericardial sac is urgently indicated

The only other differential diagnostic problem to be considered is the distinction between constrictive pericarditis and diffuse myocardial fibrosis (secondary to myocarditis or associated with endocardial fibro elastosis) These two conditions have in common (1) moderate cardiac enlargement without significant murmur (2) chronic severe right sided congestive failure and (3) catheterization data revealing (*a*) narrow pulse pressure in the right ventricle and pulmonary artery (*b*) character istic right ventricular pressure contour (Fig 159) and (*c*) high pul

monary capillary pressure Only by extensive surgical exploration can these two conditions be distinguished definitely, but because of the fact that one seldom sees a patient less than ten years of age with constrictive pericarditis, whereas a number of cases of so called constrictive myo cardial disease occur in children the pediatrician usually favors the diag nosis of constrictive myocardial disease

SPECIFIC PERICARDIAL DISEASE ENTITIES

Rheumatic Pericarditis

As mentioned before rheumatic pericarditis is the commonest form of pericarditis in children it is invariably accompanied by many of the manifestations of acute rheumatic fever I have never seen a patient with rheumatic pericarditis who did not at the same time have a loud apical systolic murmur Consequently if a patient with pericarditis has no significant murmur a diagnosis other than rheumatic fever must be considered Rheumatic pericarditis is usually of the fibrinous or sero fibrinous variety and it is characterized more by pain and friction rub than by large fluid accumulation As a rule paracentesis is not advocated in patients with rheumatic pericarditis unless tamponade is imminent The diagnosis is usually clear enough and antirheumatic treatment is successful today in a high percentage of cases The rare instance of cardiac tamponade however should be treated by paracentesis or thora cotomy This represents a real cardiac emergency and early decisive action may be lifesaving Much less harm is done by an occasional un necessary pericardial exploration than by neglecting to tap the peri cardium of a patient with true tamponade

Uremic Pericarditis

In the later stages of chronic glomerulonephritis uremic pericarditis occurs It is a manifestation of the chemical irritation of the serous mem branes by the high nonprotein nitrogen content of the plasma Usually there are no symptoms The diagnosis is based on the appearance of a friction rub in a patient dying with uremia

Purulent Pericarditis

Purulent pericarditis usually associated with bacterial pneumonia and left sided empyema is a rare but therapeutically important disease These patients show the striking combination of pericardial tamponade with the septic picture of localized accumulation of pus Chemotherapy has little if any effect on these youngsters Emptying the pericardial sac on the other hand dramatically solves both problems In this age of ex tensive chemotherapy organisms are rarely cultured from the exudate but the older literature mentions pneumococci streptococci and staphy

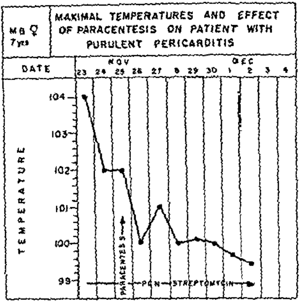


Figure 160 Temperature chart and effect of paracentesis on patient with purulent pericarditis



Figure 161 Posteroanterior radiograms of a child with purulent pericarditis (A) before and (B) after thoracotomy

lococci among the pathogenic organisms Figure 160 shows the course of a patient with purulent pericarditis Figure 161 shows the roentgenograms of this child before and after evacuation of the purulent exudate Whether constrictive pericarditis will develop later in these patients once they have been cured cannot be stated with certainty My personal feeling based on limited experience is that constrictive disease probably does not follow in the wake of successfully treated purulent pericarditis

Idiopathic or Viral Pericarditis (Acute Benign)

This is a relatively recently described disease.[8, 78] The initial case reports concerned young adults principally, but within the last few years children with this condition have also been described.[1]

The disease usually follows an upper respiratory tract infection. Occasionally it occurs in families or communities where there is an epidemic of pleurodynia. The first symptom is fever with pericardial pain and dyspnea. At physical examination these patients may appear very sick, sitting up in bed, short of breath, anxious looking, leaning forward, perhaps even clutching a pillow against the chest. On auscultation a friction rub without murmur is heard and the heart is enlarged. Electrocardiograms characteristic of pericarditis are obtained in almost every case. The sedimentation rate is increased and leukocytosis is common, but real cardiac tamponade is rare.

The outcome in all the reported cases was favorable. The illness lasts for several weeks. Chemotherapy by and large is not effective, although Aureomycin has been claimed to exert beneficial influence. Aspirin, oxygen and occasionally morphine give satisfactory symptomatic relief. The use of steroids, as has been advocated,[1, 5] can hardly be justified in a disease with such an unquestionably benign course.

Although the cessation of the individual attack within a matter of a few weeks can safely be predicted, the disease commonly recurs within a few weeks, months or even years. I have seen a boy who had three attacks of idiopathic pericarditis within two years. For recurrent attacks the prophylactic use of Aureomycin can be advocated, perhaps more on an emotional than an intellectual basis.

The differential diagnosis should be directed principally toward excluding rheumatic fever. Only secondarily should other causes of pericarditis be considered and identified by appropriate means.

Pericarditis Associated with Rheumatoid Arthritis

Pericarditis associated with rheumatoid arthritis or Still's disease should be mentioned as one of the rare cardiac complications of this syndrome. Other circulatory disturbances possibly associated with rheumatoid arthritis are (1) aortic regurgitation, (2) myocardial disease and (3) pulmonary vascular obstruction.

The clinical findings are the same as those listed in the general description of pericarditis. Tamponade is rare; the incidental discovery of a pericardial friction rub is often the only sign of pericardial involvement in rheumatoid arthritis. The outcome depends on the prognosis of the associated disease. Steroid therapy may be indicated for the sickest of these children.

Tuberculous Pericarditis

Tuberculous pericarditis is a disease of old people, but it has been noted in children occasionally.[7] The pericardial involvement is always

the result of a direct spread of the infection from the adjoining lung field. It consists of serofibrinous or hemorrhagic exudate followed by the development of granulation tissue and extensive scarring. A considerable body of opinion attributes the presence of constrictive pericarditis to long standing tuberculous involvement.[1-64]

Until recently the prognosis was very poor. With the newer chemotherapeutic advances in the treatment of tuberculosis it has improved a great deal. Tamponade is rare and the pericardial involvement is only a small part of the generalized severe tuberculosis.

NEOPLASMS

Neoplasms such as lymphosarcoma, Hodgkin's disease or primary carcinoma of the lung may invade the heart and give rise to a hemorrhagic exudate. This is usually a terminal event, and the involvement of the pericardium does not influence the clinical picture appreciably.

PERICARDIAL CYSTS

Pericardial cysts discovered on routine chest roentgenograms and commonly giving no symptoms whatsoever have been seen occasionally at The Children's Hospital. The tremendously enlarged heart, giving no symptoms and accompanied by no circulatory involvement and a perfectly normal electrocardiogram, presents a striking picture.

The radiologic and electrocardiographic findings in a boy with a pericardial cyst, which was successfully removed, are presented in Figures 162 and 163. Apparently most of these cysts are benign, and surgical removal is easy and is followed by complete recovery.

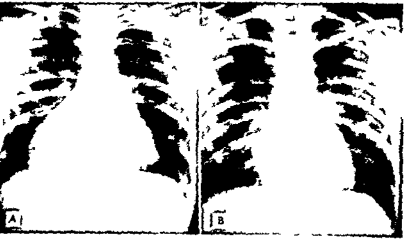


Figure 162 Posteroanterior radiograms of a boy with a pericardial cyst. (A) Before and (B) after removal of the mucilaginous structure.

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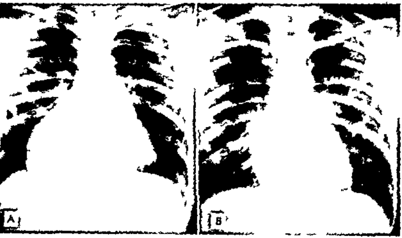


Figure 162 Posteroanterior radiograms of a boy with a pericardial cyst. *A* Before and (*B*) after removal of the mucilaginous structure

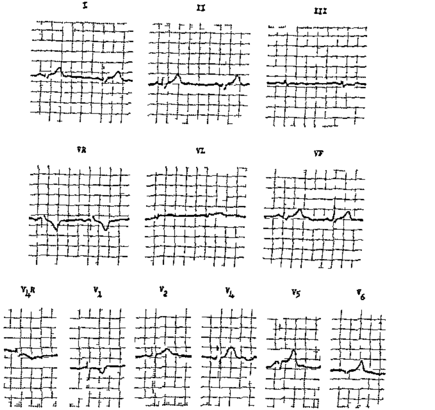


Figure 163 Electrocardiogram of a patient with pericardial cyst Note the entirely normal tracing

Chronic Constrictive Pericarditis

Constrictive pericarditis must always be considered in the differential diagnosis of chronic congestive failure of unknown etiology We have seen only a few cases of this disease at The Children s Hospital in Boston but we have had ample opportunity to see adults with this crippling condition in hospitals adjoining our institution The youngest patient with constrictive pericarditis described by White was ten years old [354] but Burwell reported the disease in children even younger than this [65]

The etiology of the condition is not known in at least half of the cases Tuberculosis is considered to be the underlying disease in the other 50 per cent The physiologic handicaps of chronic constrictive pericarditis are identical to those described in cardiac tamponade and summarized in Table 17 (p 256) At cardiac catheterization high venous pressure and high right atrial pressure are found There is usually an early diastolic dip in the right ventricular pressure curve followed by a high end diastolic pressure (Fig 159) Pulmonary arterial mean and pulmonary capillary pressures are elevated and the cardiac output is diminished

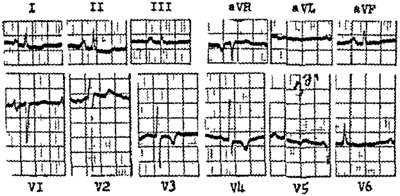


Figure 164 Electrocardiogram of patient with constrictive pericarditis

The clinical picture is characterized by the triad of a small heart elevated venous pressure and ascites The ascites is out of all proportion *to the edema elsewhere The patients are not as sick as one would expect* from the marked evidence of right sided failure they may be able to do a moderate amount of exercise and orthopnea is rare

The physical examination is dominated by the consequences of high right atrial pressure and low cardiac output Paradoxical pulse is noted and the pulse pressure is narrow The heart sounds may be somewhat diminished no significant murmurs are audible and the rhythm except in older people is of sinus origin A third heart sound at the lower left sternal border is relatively common but of no particular diagnostic significance in the younger patients

Radiologic examination reveals a heart that is only moderately if at all enlarged this is in striking contrast to the marked congestive failure Calcification in the pericardium is pathognomonic and occurs in as many as 55 per cent of the patients in some series [79] At fluoroscopy the cardiac beat is diminished sometimes more on the right than on the left side The electrocardiogram shows low voltage T wave inversion and a broad notched P wave (Fig 164)

Constrictive pericarditis if it leads to the clinical syndrome here described should be treated surgically Radical pericardiectomy as advocated by Holman and his associates [169] is the preferred approach to the problem This consists in decortication of the pericardium beyond the left right and inferior cardiac borders including liberation of the venae cavae The results were good in fourteen of the twenty six patients in Holmans latest series Four of the twenty six died Improvement can be measured largely in terms of a decrease in venous pressure and increase in cardiac output

Congestive Failure

Definition and Physiology

Congestive failure in the light of newer knowledge concerning circulatory physiology is increasingly hard to define Lewis [99] regarded cardiac failure as "inability of the heart to discharge its contents adequately This was a perfectly good definition until it was recognized that in many instances of chronic congestive failure the ventricle does fulfill its obligation but is able to do so only by invoking compensatory mechanisms such as tachycardia increased diastolic filling pressure or cardiac hypertrophy Lewis classical definition was further qualified when on the basis of animal experimentation it became clear that relatively minor impairments of cardiac function may be reflected solely in the decreased capacity of the kidneys to excrete large salt loads [7]

Since at present no precise physiologic definition is available, it might be well to regard congestive failure simply as a clinical syndrome associated with heart disease Left sided failure is characterized by pulmonary rales accentuated pulmonary closure gallop rhythm left hydrothorax pulsus alternans dyspnea orthopnea and cardiac asthma

In contrast right sided failure is identified by increasing systemic venous pressure enlargement of the liver dependent edema right hydrothorax hydropericardium and oliguria

Cardiac enlargement is present in both left sided and right sided failure cyanosis may or may not be present in either

The congestive failure syndrome is frequently encountered in pediatric hospitals the age distribution is of some interest In patients with congenital heart disease failure is most likely to develop within the first six months of life if this period is survived without evidences of congestion it is not likely to occur until at least two years of age or even considerably later On the other hand congestive failure associated with rheumatic heart disease usually does not occur before the age of four to five years and it always indicates an active rheumatic process Paroxysmal atrial tachycardia and primary myocardial disease—two relatively common causes of congestive failure in childhood—usually manifest themselves within the first year or two of life

The two most commonly quoted theories of congestive failure are the backward and forward failure concepts According to the theory of backward failure [161] the heart, consisting of two independent pumps (the

left and the right ventricles) fails when one pump cannot forward the load offered to it by the other For instance a failing left ventricle being unable to put out all the blood the right ventricle offers acts as a dam behind which the blood rapidly accumulates increasing the volume and later the pressure in the left atrium pulmonary veins and even the pulmonary artery and the right ventricle This theory would explain very well the clinical syndrome referred to as left sided failure consisting principally of pulmonary congestion and its consequences The reason this explanation holds so well for left sided failure is the rather limited capacity of the pulmonary vascular bed where even a few hundred extra cubic centimeters of blood may cause considerable trouble

If on the other hand one tries to explain the phenomena observed in right sided failure that is systemic congestion on the basis of backward failure alone one encounters a considerable amount of difficulty The principal obstacle to this explanation for failure of the right ventricle is the fact that even if the entire contents of the pulmonary circuit—roughly one tenth of the circulating blood volume—were transferred to the systemic circuit this by itself should not cause any trouble the systemic reservoir is so large that simply shifting some (or even all) of the blood from the lungs should not cause an elevated systemic venous pressure any more than a good sized blood transfusion would Furthermore ligation of the inferior vena cava performed therapeutically as well as experimentally does not produce evidences of systemic congestion with any regularity [9] Finally complete destruction of the right ventricle in animal experiments is tolerated relatively well, and certainly without evidences of failure [3 9]

Proponents of the forward failure theory[241] maintain that the congestive failure syndrome is due to a low cardiac output resulting in an inadequate supply of oxygen to the tissues increased capillary permeability and renal failure This then would give rise to increased blood volume edema and other manifestations of congestive failure The most obvious difficulty in accepting this theory is the observation that severe anoxic states such as cyanotic congenital heart disease may exist for many years without any evidence of congestion furthermore no really good evidence of increased capillary permeability in cardiac failure exists nor is there a straight line correlation between the absolute level of cardiac output and the presence or absence of failure

Since neither of these two theories alone explains the phenomena described perhaps a combination of several factors may result in a more acceptable hypothesis Burch [58] Altschule[10] and others[1 3] have proposed theories that fit in broadly with the concepts to be presented The entire thought however is based on Starling s[319] fundamental work on the heart lung preparation

Perhaps the outstanding feature of congestive failure is the failure of the cardiac output to meet all the demands placed on it by the metab

olism of the body At first this may manifest itself only if the demands are significantly increased such as with severe exercise Later the demands may not be met even at rest and finally the low cardiac output becomes incompatible with life itself

When the cardiac output becomes relatively inadequate the first mechanism that comes into play is a reflex increase of the heart rate Thus even with an unsatisfactory stroke volume the cardiac output per minute may be adequate This is an inefficient type of compensation however since the tachycardia by and large is achieved by shortening diastole the resting period of the cycle during which the coronary arteries fill

A more efficient way the heart can try to increase its lagging stroke volume is by increasing the end diastolic pressure in the ventricles this is achieved by raising atrial and venous pressures It has been pointed out before that this may be achieved easily in regard to the pulmonary veins and the left atrium by merely shifting the blood volume In order to increase the pressure in the systemic veins and the right atrium however the increase in systemic blood volume alone is not enough One must assume the presence of a reflex increase in the tone of the veins as well The increase in blood volume is the result of sodium and water retention a renal component of cardiac failure possibly negotiated through the antidiuretic hormone of the pituitary It should be pointed out here that the cardiac output—or stroke volume—achieved by the ventricle at the price of this increase in ventricular diastolic and venous pressures is not commensurate with the output achieved by a corresponding increase in venous pressure in a normal heart The failing heart is an inefficient motor needing the artificial stimulus of increased venous pressure to achieve average or normal cardiac output The continuous increase in diastolic filling results first in dilatation of the ventricle and later gives rise to hypertrophy The hypertrophied heart is reputedly a more efficient organ than the dilated one although good experimental evidence to this effect is missing

Through these mechanisms of tachycardia increased diastolic pressure and myocardial hypertrophy the failing heart can maintain compensation that is it is able to achieve a more or less satisfactory cardiac output for a while As the condition progresses however cardiac efficiency becomes less and less adequate The amount of work performed per cubic centimeter of oxygen consumed diminishes Higher and higher venous pressure is needed to put out relatively small stroke volumes Finally a point is reached at which the stroke volume actually declines with further increase in venous pressure The heart muscle has arrived at the declining part of Starling s curve compensation cannot be further maintained the cardiac output becomes inadequate for the maintenance of vital functions and death ensues

CLINICAL PICTURE

How do the phenomena just discussed explain the clinical features of congestive failure?

Venous hypertension is the cardinal sign of right sided failure Its significance and measurement have been described previously (p 17)

Edema is probably a consequence of the increased venous pressure overpowering the normal or lowered oncotic pressure of the plasma proteins Since the lymph flow is also hindered by the increased venous pressure probably there is not only an increased production of interstitial fluid but also a slower removal through the thoracic duct Among the sites of edema in children with right sided congestive failure facial edema must be mentioned first, and enlargement of the liver next Ascites and anasarca are relatively late manifestations and they usually occur after considerable enlargement of the liver has been noted Hydrothorax and hydropericardium are rare

Dyspnea is a subjective phenomenon thus not subject to quantitative appraisal *Hyperpnea* on the other hand can be measured with ease Hyperpnea in congestive failure is probably caused by the stimulation of a number of complex chemoreceptors and pressure receptors in the systemic and pulmonary arterial trees as well as in the medulla The stimuli acting on these receptors may be anoxia hypercapnia increased lactic acid content of the blood pulmonary rigidity or congestion

Cyanosis in congestive failure may be due either to decreased oxygen intake through congested lungs or to increased deoxygenation by the oxygen hungry tissues during a prolonged circulation time in the dilated capillaries

Orthopnea and pulmonary edema can best be explained by left ventricular failure accentuated by the increased pulmonary blood volume and diminished vital capacity inherent in the horizontal position Pulmonary rales are evidences of fluid accumulation in the lungs

Pulsus alternans (see Fig 7 p 17) of the systemic arteries—an evidence of left ventricular failure—can best be detected when the patient s blood pressure is being taken As the sphygmomanometer is slowly released at one point every second arterial beat will come through the stethoscope As the pressure in the manometer is dropped another 5 to 10 mm the number of sounds heard per minute suddenly doubles and becomes equal to the apex beat It is believed that the alternation is due to the weakness of the individual cardiac fibers resulting in the alternately slower and weaker contractions

Within recent years pulsus alternans of the pulmonary arterial system has been noted at cardiac catheterization in patients with right ventricular failure [118]

X ray evidence of cardiac enlargement is almost a sine qua non of the diagnosis of congestive failure Usually the enlargement is generalized, involving all chambers At fluoroscopy the cardiac beat is poor

The enlargement and the poor beat are thought to be due to the in creased diastolic volume and the poor stroke output In left sided failure pulmonary congestion can be easily detected by roentgenogram

Electrocardiographic evidences of congestive failure are furnished by the presence of P waves suggesting atrial hypertension

The classic picture of congestive failure in children is one of right sided or combined right and left sided failure dramatic evidences of pure left sided failure such as pulsus alternans or orthopnea are rarely observed However pulmonary rales can be heard in at least half of the patients with frank right sided failure and gallop rhythm is commonly encountered

Right sided failure in its pure form may be seen in the following conditions pulmonary vascular obstruction pulmonary valvular obstruc tion tricuspid valve disease atrial septal defect pulmonary venous anomaly and various types of cyanotic congenital heart diseases (trans position aortic atresia and so forth)

Pure left sided failure may be observed in severe systemic hyper tension (renal disease) coarctation of the aorta aortic stenosis mitral valve obstruction and patent ductus arteriosus

A combination of left sided and right sided failure with the sys temic congestion dominating is the commonest form of congestive fail ure in children All the conditions which may cause pure left sided fail ure may also cause combined failure In addition ventricular septal de fect single ventricle and truncus arteriosus should be mentioned among the congenital conditions which may cause combined failure For the most part the diseases in this group and all conditions causing conges tive failure are those which basically involve both ventricles such as rheumatic fever primary myocardial disease paroxysmal atrial tachy cardia and acute glomerulonephritis Constrictive pericarditis a rare condition in youngsters also results in combined ventricular failure

Treatment

Before the advent of cardiac surgery and the various therapeutic and prophylactic measures directed against rheumatic fever treatment of congestive failure was about all one could do for children with heart disease Obviously the picture is different today still the successful management of congestive failure is one of the severest tests of the skill of the pediatrician

Symptomatic Treatment When the pediatrician sees a patient with congestive failure his first task is to make the child more comfortable Usually sedation—morphine or even phenobarbital—is indicated since many of the older children are fretful apprehensive and frightened and the babies are irritable Sedation need not be continued for a long time but a little morphine sulfate for the first day or two gives much needed rest and invaluable support to the patient It also lessens the impact of

the various and inevitable therapeutic or diagnostic procedures to which the patient is subjected when his disease is first discovered

Although the therapeutic value of oxygen in congestive failure has not been proved conclusively the symptomatic relief afforded by it is unquestionable Babies if they are too small to worry about an oxygen tent and children if their fears have been alleviated by sedation and appropriate explanation will relax breathe easier and act a great deal happier in an oxygen tent with about 50 per cent oxygen concentration Nasal oxygen is bothersome and inefficient and should be used only if a tent is not available Since the prime purpose of oxygen administration is to make the child *more comfortable* it does not make sense to insist on its administration for a child who is terrified of the tent I feel strongly that unless the child can be sedated or reassured to a sufficient degree the use of a tent by causing terror and apprehension may actually be harmful

Decreasing the Load of the Heart If the assumption that congestive failure is connected somehow with inadequate cardiac output is correct then it is desirable to decrease the demands made on the circulation

One way to accomplish this is by keeping the patient in bed For adults and older children this is the procedure of choice in almost all instances in small babies the problem does not arise Toddlers and young children however may need special consideration If one realizes that bed rest is desirable only as a means of achieving as much rest as possible then it becomes obvious that if a child is upset and frustrated by being kept in bed this therapeutic maneuver much like the oxygen tent under certain circumstances defeats its own purpose In other words rest should be interpreted as broadly as possible Most children with failure actually welcome being put to bed On the other hand some overactive youngsters rest better by sitting in a high chair being read to looking at television or playing in a play pen The doctor s individual judgment about each patient should supersede here—as in most medical decisions—any preconceived ideas obtained from dogmatic textbooks

If arterial hypertension of significant degree is present as in renal disease lowering of the blood pressure by hypotensive drugs is indicated

Decreasing the load on the kidneys in congestive failure may be achieved by restricting the sodium intake to 500 mg or less per day This may be accomplished by not adding salt to the food by using milk with a low sodium content (Lanolac) and by avoiding heavily salted foods (bacon and so forth) Water may be taken ad libitum as long as the salt intake is controlled

Drugs DIGITALIS As a matter of principle every child with obvious evidence of congestive failure should be digitalized The only possible exceptions to this general rule may be patients whose cardiac failure is caused by pericardial tamponade or anemia Even in these two situations digitalization probably does not do any great harm The only

true contraindication to the use of digitalis in congestive failure is the presence of digitoxicity

The effect of digitalis on the heart has been discussed by internists pharmacologists and physiologists for many years[139] [1] The use of this glycoside in pediatrics has recently been summarized Digitalis causes an increase in myocardial responsiveness an increase in contractile force a slowing of the pulse rate a decrease in venous pressure and shortening of the circulation time

The digitalis preparation to be used depends a great deal on the experience of the individual physician All digitalis preparations put out by reputable drug companies today can be used effectively and safely The difficulty arises from the multiplicity of drugs with similar names (almost all using the dig prefix in some fashion) and radically different absorption and excretion patterns as well as toxic effects Every doctor should become familiar with one rapidly acting parenteral preparation and one slowly or moderately rapidly acting drug that can be used orally as well as parenterally The frequent and judicious use of only a few preparations enables the pediatrician to learn by personal experience the mode of action of the drug its toxic manifestations and its therapeutic efficacy It is hard to resist the lure of advertisements flooding the medical periodicals with a newer and better drug every month, each one supported by more and more learned quotations from the literature In order to preserve one s own sanity as well as to promote the patient s welfare one should reject these wonderful new adventures in digitalization and persist with the few drugs one has learned to work with through the years I usually use one of three drugs

Digitoxin Digitoxin is a relatively slowly acting glycoside which may be administered orally intramuscularly or intravenously The oral absorption is virtually complete hence the great advantage that oral and parenteral dosages do not differ significantly Oral preparations are available in liquid and tablet form

The maximal effect of the drug occurs four hours after intravenous administration and six to eight hours after oral dosage Most of the glycoside is excreted within ten to fourteen days but elimination may not be complete for three weeks The slow excretion of digitoxin has the disadvantage that toxicity once it occurs persists for a relatively long time the advantage is easy maintenance of digitalization

A satisfactory dosage schedule (Table 12 p 184) may be obtained if the age and the weight of the patient are taken into consideration Patients less than two years old need 0 02 to 0 03 mg per pound of body weight whereas children more than two years of age can be digitalized with 0 01 to 0 02 mg per pound This amount should be given in three divided doses within a twenty four to thirty six hour period Digitalization can be maintained by one tenth of the total digitalizing dose

Cedilanid Cedilanid is a rapidly acting parenteral drug primarily

designed for intravenous administration but it may also be given intramuscularly

Appreciable effects of the drug may be demonstrated ten to fifteen minutes after intravenous administration and maximal effects are achieved within one hour Virtually complete elimination occurs in twenty four to thirty six hours This rapid action and excretion offer great advantages in the handling of emergencies such as occur in the operating room and they make this an ideal drug to use when one can not be sure whether the patient has had too little or too much digitalis

The total digitalizing dose is about the same maybe 10 per cent more than that outlined for digitoxin (Table 12) If the patient is to be maintained for several days or weeks with intravenous injections of Cedilanid one quarter of the digitalizing dose should be given every six hours It is a much better policy however to use Cedilanid only as the initial rapid digitalizing agent and then to maintain the level of digitalization with a slowly acting drug administered orally or intramuscularly If this is to be done the patient should be cautiously redigitalized with the slower acting preparation using the lower dosage scales within twenty four to thirty six hours after the last dose of Cedilanid Maintenance doses then can be calculated on the basis of the slower acting glycoside alone

Digoxin Digoxin is a good all round preparation It may be used orally as well as parenterally in emergencies as well as in chronic situations

In terms of rapidity of absorption and excretion it stands midway between digitoxin and Cedilanid Full effects of Digoxin may be demonstrated four hours after its oral administration A significant portion of the initial dose is excreted within twenty four hours most of it disappears within forty eight to seventy two hours A debatable point has been the completeness of the absorption from the gastrointestinal tract which would obviously affect the relative size of oral and parenteral dosages My impression is that parenteral dosages should be calculated at about 50 to 75 per cent of the oral dose

The oral dose administered in liquid or tablet form is about 0 04 to 0 06 mg per pound of body weight for children less than two years of age and 0 02 to 0 04 mg per pound for those more than two years old (Table 12) Daily maintenance doses can be calculated at 25 per cent of the digitalizing dose

Effectiveness It cannot be emphasized strongly enough that the dosage schedules outlined for these three digitalis preparations can serve only as guide posts *The digitalization of every patient should be an individual experiment* For the first eighteen to twenty four hours the lower of the two dosages suggested should be administered If at that time the therapeutic goal has not been reached and there are no signs of toxicity digitalis to the amount suggested by the higher of the two figures

should be administered within the next twelve hours If toxicity or therapeutic effects still are not noted further increments should be given cautiously checking one s position every twelve hours until either the desired effect is obtained or evidences of overdosage are noted

One of the best indices of therapeutic effectiveness is slowing of the pulse A significant drop in heart rate with disappearance of gallop rhythm may often be evidence of good digitalis effect Other and equally significant therapeutic aims are a decrease in the size of the liver disappearance of pulmonary rales and edema diminishing respiratory rate and improvement of color appetite and disposition In older children weight loss is a significant index of successful digitalization In small infants however because of the relatively small absolute amounts involved and the general inaccuracy of weighing methods weight loss cannot be used as a sensitive measure of effectiveness of digitalization

In contrast to the general accuracy of clinical observations the electrocardiogram is of no use whatsoever in determining whether digitalization has been successful in the alleviation of congestive failure There is almost nothing more exasperating to an electrocardiographer than to be asked by a colleague Has this patient been fully digitalized yet? As pointed out in a previous chapter (p 56) the electrocardiogram can show only whether there is some or too much digitalis in the myocardium it is totally unable to gage whether there is enough present

Ordinarily evidences of digitoxicity may be readily obtained from the electrocardiogram In addition to the cardiographic changes digitalis toxicity may cause vomiting headache and visual disturbances Vomiting is the only common clinical manifestation of overdosage however

The response of infants and children in congestive failure to proper digitalization depends a great deal on their type of heart disease On the whole infants with cyanotic congenital heart disease respond poorly whereas patients with paroxysmal atrial tachycardia idiopathic myocarditis subendocardial fibroelastosis acute glomerulonephritis coarctation of the aorta or patent ductus arteriosus respond relatively well Patients with atrial septal defect ventricular septal defect and pulmonary stenosis may derive some benefit from the drug but no complete alleviation of the symptoms is usually noted Whether children with active rheumatic carditis respond to digitalis or not has been a subject of heated debate for a long time I believe that a patient with rheumatic heart disease and congestive failure (thus by definition having active rheumatic fever) should be treated with digitalis although the results may not always be as striking as in other conditions they may be rewarding indeed It may be worth while to mention that patients with exclusively or predominantly left ventricular failure have a better chance to respond favorably to digitalis than those in whom congestion is exclusively right sided

In our experience the patients who derive the most benefit from the glycosides are the ones who are most likely to show evidences of intoxica

tion It almost seems as though therapeusis and toxicity were related to each other In effect the children most commonly showing digitalis intoxication were the ones with definite myocardial involvement (primary myocardial disease or rheumatic heart disease) It is also interesting to note that these patients had received relatively small doses of digitalis when they showed evidences of toxicity Most of the toxic effects were electrocardiographic in nature and have been discussed before (p 89) Vomiting occurred rarely and only in patients receiving large doses (more than 0 03 mg of digitoxin per pound of body weight)

Treatment of Digitalis Intoxication Treatment of digitalis intoxication consists in immediate withdrawal of the drug If the atrial arrhythmia resulting from digitoxicity is severe enough to warrant treatment (paroxysmal atrial tachycardia with block) oral administration of potassium salts (potassium chloride) in doses of 1 to 2 gm per day is indicated In the rare instance of ventricular arrhythmia in children Pronestyl may be used in addition to potassium chloride

Diuretics Diuretics are indicated if digitalis bed rest and sodium restriction do not quickly and effectively control congestive failure In markedly edematous children it may even be desirable to start the administration of a diuretic simultaneously with the administration of digitalis As a rule however principally for methodical reasons it is wise to administer digitalis first and give the diuretic only if necessary

In the modern doctor s therapeutic armamentarium diuretics are practically synonymous with mercurial diuretics Although occasionally acidifying salts (ammonium chloride) and xanthine derivatives (Theocalcin aminophylline) are used as diuretics by themselves in most instances they are given only as adjuvants with mercurials

The effect of mercury is directed principally toward the kidney the site of action is most likely the distal end of the convoluted tubules Here by means of enzymatic inhibitory action mercury slows down the reabsorption of electrolytes (chloride and sodium) and water

Since water diuresis is probably secondary to a negative electrolyte balance and since tubular function is dependent directly on glomerular filtration and renal plasma flow it is necessary that the glomeruli offer the tubules a sufficient amount of plasma of average electrolyte content to ensure a good mercurial diuresis It follows then that if the plasma chloride level is low administration of mercury may not result in satisfactory diuresis[31] Correction of the plasma chloride level makes these patients mercury sensitive People with poor cardiac output and poor renal plasma flow may become equally resistant to mercury a condition sometime easily corrected with digitalization Finally in some "mercury resistant patients good diuresis occurs if the administration of the diuretic is followed half an hour or an hour later by an intravenous dose of aminophylline improving the glomerular filtration rate Within recent years a carbonic anhydrase inhibitor (Diamox) has also been used

to improve mercurial diuresis [9] Finally, the skillful use of ammonium chloride also may enhance the effectiveness of mercurial diuretics

The two mercurial preparations which I use most frequently are Mercuhydrin for intramuscular and Thiomerin for subcutaneous administration For practical purposes mercury should never be given intravenously Sudden deaths have been reported after administration of the drug in this fashion

The amount of mercury to be given (Table 12 p 184) is the minimal dose achieving the desired effect the dose of Mercuhydrin and Thiomerin is about the same One should start with 0 1 cc as a test dose to be followed the next day by a dose of 0 25 cc in infants and children less than four years old and 0 5 cc in older patients If this dose results in effective diuresis it should be repeated every two to three days if it is not effective the dose may be increased by 0 25 cc increments until a maximum of 1 cc is reached in the younger ones and 1 5 cc in the older children

Injections of mercury should be continued until the patient reaches his so called dry weight that is a weight without visible edema and without further weight loss after continued administration of the same doses of mercury

The amount of mercury necessary to keep the patient at this dry weight varies considerably and can be determined only by accurate daily weighings Some patients once they have reached the dry weight level may be kept there indefinitely with only digitalis and a low salt diet Others may need mercury once a week while still others may have to have it two or three times a week Accurate charting of the patient s weight is the only way to critically assess the need for diuretics Attempts to measure the urinary output and fluid intake particularly in small children are useless inaccurate and time consuming

Mercury should almost never be given more often than every second day because of the possible cumulative effect on the renal parenchyma Prolonged administration of mercury should be accompanied by frequent determinations of the plasma electrolyte levels It is easy to see how prolonged electrolyte depletion of the organism by diuretics accompanied by a limited salt and liberal water intake may lead to severe electrolyte disturbance the so called low salt syndrome The relationship of potassium depletion to digitoxicity has been alluded to previously [21]

To counteract the electrolyte depleting effect of the diuretics and to potentiate its effectiveness intermittent administration of ammonium chloride for two day periods preceding the administration of mercury has been recommended The average dose for children is between 1 and 1.5 gm daily To avoid digitoxicity by maintaining a satisfactory plasma potassium level continuous high potassium intake (either a quart of orange juice or 1 to 1 5 gm of potassium chloride daily) is recommended if mercury is to be used for a long time As mentioned before serum

chemistry determinations every week or every two weeks are necessary with an aggressive plan of treatment of this sort

Aminophylline in doses of 0 1 to 0 2 mg may increase the effectiveness of mercurials presumably by increasing the glomerular filtration rate and the renal plasma flow Diamox in doses of 50 to 250 mg per day [93] given for forty eight hours prior to administration of mercury may result in increased mercurial diuresis by causing hyperchloremic acidosis

Severe toxic effects of mercury are rare with intramuscular administration Chills fever rashes and arrhythmias have been reported These may be avoided by using the principle of the test dose before administration of therapeutic doses I know of one patient who was recovering from bacterial endocarditis in whom a fever developed a few hours after intramuscular administration of Mercuhydrin Subsequent skin tests did not show any allergic reaction Consequently a few days later mercury was again given intramuscularly Again it was followed in a few hours by fever and this time death Whether this patient suffering from severe congenital heart disease and recovering from bacterial endocarditis died as a result of mercury intoxication cannot be proved but such a possibility exists Several instances of sudden death minutes after intravenous administration of the drug have been reported

Mercury should not be given to patients with obvious renal disease either acute or chronic It is a good rule of thumb never to use mercurials if there is a nonprotein nitrogen level of more than 50 mg per 100 ml of blood

Treatment of the Underlying Disease As mentioned previously at least in terms of pediatric cardiology the concept that congestive failure could be treated by attacking the underlying disease rather than by alleviating symptoms is only ten years old

The first things to be mentioned in this regard are the miracles of cardiac surgery! Mitral stenosis aortic stenosis constrictive pericarditis congenital arteriovenous aneurysms patent ductus arteriosus pure pulmonary stenosis atrial septal defect and even coarctation of the aorta all may result in congestive failure in children and all can be treated surgically Of course surgery under these circumstances is fraught with more than the usual amount of danger nevertheless the risk is obviously *well worth taking The dangers of surgery may be considerably lessened* by intelligent anticongestive treatment preoperatively in this regard however one should be careful to use a quickly eliminated digitalis preparation so that the cardiac hyperexcitability caused by digitalis will not predispose to arrhythmias at the time of operation It may be best to stop digitalis therapy for forty eight hours before operation It is equally important to ensure as far as possible an optimal electrolyte profile in the preoperative and postoperative periods

Another disease causing congestive failure in children for which

etiologic treatment may be available is acute rheumatic fever The pros and cons of steroid therapy have already been outlined and the tentative indications and dosages have been proposed (p 143) Occasionally in a case reported in the literature it is suggested that hormone therapy may have its place in the management of the primary myocardial disease entities as well

Thyrotoxicosis seldom causes congestive failure in children If it does a course of thiouracil therapy followed by surgery is the choice of etiologic treatment

Congestive failure associated with anemia can of course be corrected by increasing the circulating hemoglobin level I am certain that in a book written primarily for pediatricians the importance of slow and gradual transfusions for severely anemic patients does not need to be stressed These patients have too little hemoglobin and too much blood volume hence small amounts (50 to 100 cc at a time) of packed cells should be given several times twelve to twenty four hours apart rather than one large transfusion which may be just too much for a heart already under maximal stress

CHAPTER 13

Miscellaneous Diseases

HYPERTENSIVE HEART DISEASE

ETIOLOGY

Essential hypertension the principal cause of persistently elevated blood pressure in adults is rare in children Renal disease diseases of the central nervous system coarctation of the aorta and endocrine disturbances are more common etiologic factors [147, 31] The technique of obtaining accurate blood pressure readings and the normal values for various ages have already been discussed (see p 14) Table 17 lists the most important factors causing high blood pressure in children with appropriate diagnostic tests for their identification

Renal Disease The most important cause of elevated blood pressure in children is renal disease In addition to the laboratory tests enumerated in Table 17 a history of kidney disease is an important feature in the diagnosis even when results of urinalysis and renal function tests are normal

Diseases of the Central Nervous System As outlined in Table 17 diseases of the central nervous system can usually be diagnosed by the clinical picture and results of laboratory tests including studies on the cerebrospinal fluid roentgenograms of the skull and electroencephalograms Pneumoencephalograms are rarely necessary The syndrome of familial dysautonomia [90] can usually be recognized by the labile hypertension absence of tearing repeated febrile episodes hypesthesia motor disturbances and mental retardation

Cardiovascular Diseases The cardiovascular diseases listed in Table 17 have been amply discussed in appropriate chapters

Diseases of the Endocrine System The most frequent endocrine cause of hypertension today is prolonged administration of cortisone Obviously the clues to this diagnosis are a history of steroid administration and the appearance of Cushing's syndrome

Pheochromocytoma is difficult to diagnose it is also extremely rare in children only sixteen cases having been reported in the literature [93] At The Children's Medical Center only one authenticated case has been found The classic picture caused by an increase in the amount of epinephrine in the blood consists in paroxysmal hypertension intermittent sweating dilated pupils headaches nausea vomiting tachy

Table 17 Hypertension in Infants and Children Causes and Diagnostic Aids

Etiology	*Diagnostic Aids*
I Renal	
A Acute and chronic glomerulo nephritis	Clinical picture
	Repeated urinalyses Addis count
Chronic nephritis secondary to anaphylactoid purpura	Concentration test NPN
	Urea and creatinine clearance
	Renal biopsy
B Pyelonephritis	Repeated urinalyses
	Repeated urine cultures plus above tests in chronic cases
C Polycystic renal disease	Intravenous pyelogram
D Tumors Wilms and neuro blastoma	Retrograde pyelogram
	Split renal functions
	Exploratory laparotomy and renal biopsy
E Ectopic kidney	
F Hypoplastic kidney	
G Hydronephrosis	
H Renal artery and vein anomalies and thromboses	Above tests plus aortogram and split renal functions
I Polyarteritis disseminated lupus erythematosus	Clinical and laboratory signs of vascular disease plus renal biopsy
J Primary arteriolar disease of the kidney	Azotemia urine sediment abnormalities
	Renal biopsy or necropsy
II Central nervous system	
A Poliomyelitis	Clinical picture
	Lumbar puncture
B Encephalitis	Above plus electroencephalogram
C Brain tumor	Skull x rays
	Above plus pneumoencephalogram or ventriculogram
III Cardiovascular	
A Coarctation of aorta	Femoral pulses (weak compared with radial)
	Blood pressure in legs
	X ray picture (collateral esophagogram)
B Patent ductus arteriosus	Continuous murmur 2nd LIC
	EKG left ventricular hypertrophy
	Cardiac fluoroscopy
	Rarely retrograde aortogram
IV Endocrine	
A Secondary to cortisone or ACTH therapy	History
B Pheochromocytoma	History IVP
	Regitene Benzodioxane tests
	Histamine test
	Urinary or blood catechol amine
C Cushing s syndrome	Clinical picture
	Endocrine studies
D Congenital adrenal hyperplasia	Clinical picture
	17 ketosteroid excretion

V Poisonings	
A Lead	Clinical picture
	Anemia stippled cells
	Cerebrospinal fluid (elevated pressure and protein)
	Glycosuria
	X ray of long bones and skull
	Urine coproporphyrin (qualitative)
	Urine lead excretion (quantitative)
B Mercury	Clinical picture of acrodynia
	Urine mercury excretion
VI Essential	Positive family history
	? Cold pressor and amytal tests

cardia and pallor Occasionally an abdominal mass may be palpated compression of which may induce the symptoms described Among the specific tests mentioned in Table 17 an increase in urinary catechol amines is the most reliable one Intravenous pyelography which may reveal a mass in the region of the kidney is perhaps the easiest and most helpful of the clinical laboratory tests even though it does not demonstrate those pheochromocytomas lying outside the adrenal area (approximately 12 per cent) The regitine and benzodioxane tests are useful if the patient has significant hypertension at the time of testing an appreciable drop in systolic blood pressure may occur when one of these drugs is administered[135 138]

Poisonings Poisonings that produce hypertension can usually be excluded on the basis of the history the physical examination and the laboratory tests indicated in Table 17

Essential Hypertension Essential hypertension is principally a diagnosis of exclusion if a child with significant hypertension does not have any of the conditions enumerated so far one must assume that his hypertension is essential No specific diagnostic test for essential hypertension exists

As mentioned previously this is a rare disease in children we have seen nine patients with this disease less than fourteen years of age at The Children s Medical Center[137] A critical evaluation of the English and American literature revealed only another ten well authenticated cases

The ages of our patients ranged from seven months to thirteen years Five of these children had severe hypertension (pressures consistently higher than 160/110) and secondary circulatory changes (cardiac enlargement left ventricular hypertrophy and hypertensive retinopathy) The blood pressure of the other four patients was never found to be higher than 160 110 No secondary circulatory changes were present All the members of the group with mild hypertension had a family history of essential hypertension whereas only one in the group

of five with marked elevation of the blood pressure had a family history of such The disease in the former group may well be related to essential hypertension in adults whereas the severe hypertension in the latter group may represent a completely different etiologic entity

The prognosis of the mildly hypertensive group throughout childhood is good On the other hand two of our five patients with severe hypertension died

Treatment

Obviously the individual entities within the general hypertensive group should receive etiologic treatment if possible In patients with unilateral renal disease removal of the affected kidney may give miraculous therapeutic results Among patients with primary disease of the central nervous system the hypertension either may subside with the disappearance of the underlying inflammatory disease or may be cured by successful removal of a neoplasm Surgical treatment of the congenital heart diseases causing hypertension can effect a cure relatively easily Cessation of ACTH or cortisone therapy or removal of a pheochromocytoma may permanently lower the elevated blood pressures caused by these factors Elimination of the poisons from the environment obviously will relieve hypertension caused by ingestion of mercury or lead

The treatment of essential hypertension is as difficult a problem in children as it is in adults Mild sedation weight reduction and sustaining psychotherapy have been and still are the mainstays of treatment Moderate physical activity is not only harmless but even beneficial in many instances

If these relatively simple measures do not control hypertension drug therapy should be considered for its immediate therapeutic and future prophylactic effects Table 12 (p 184) lists the various drugs available their dosages toxic effects and indications for their use At first, in our clinic patients with mild hypertension are usually given Rauwolfia serpentina in doses indicated in Table 12 Every two to three weeks the dose is increased by ½ to 1 tablet a day until either the desired *hypotensive* effects are achieved or prohibitive side effects (drowsiness nausea diarrhea or nasal stuffiness) occur If side effects appear the dose is slightly reduced and the patient is maintained on this dosage If no significant reduction of blood pressure is achieved after several weeks Apresoline (hydralazine) is usually added in appropriate doses to the Rauwolfia The dosage is gradually increased until it reaches 300 to 500 mg of Apresoline per day

If a combination of Rauwolfia and Apresoline does not achieve the desired purpose hexamethonium is added to the previous drugs as may be seen in Figure 165 This drug often significantly lowers the blood pressure in patients in whom the other two alone had had no effect Patients taking hexamethonium must be warned concerning constipa

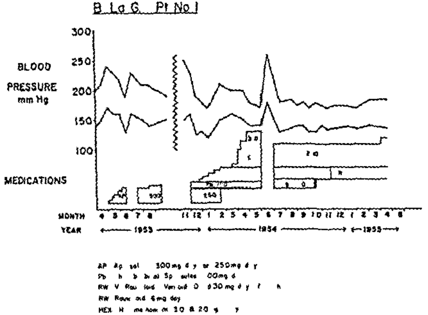


Figure 165 Response of essential hypertension to a combined treatment with Rau wolfia Apresoline and hexamethonium

tion urinary retention and postural hypotension At the first sign of any of these the dosage should be reduced and cathartics should be given

For patients with severe essential hypertension who are not bene fited by any of these drugs sympathectomy is recommended Four of our five patients with severe hypertension had this formidable surgical procedure in two cases the results were good but in the other two there was no change in blood pressure At present it is impossible to predict which patient will or will not respond to sympathectomy

One further word about the treatment of patients with so called hypertensive encephalopathy such as is seen in acute and chronic glomerulonephritis chronic poliomyelitis or coarctation of the aorta The therapy of these patients taxes the ingenuity of the pediatrician Perhaps the most useful drug to start with is magnesium sulfate in con trolled intravenous drip (Table 12) 0 1 cc per pound of body weight of a 3 per cent solution but no more than 75 to 100 mg per hour If this does not reduce the pressure protoveratrine should be administered intravenously with extreme caution starting with 0 5 to 1 microgram per pound of body weight and increasing the dose by minute amounts every thirty minutes If the correct intravenous dose is determined one can switch to intramuscular administration of protoveratrine or mag nesium sulfate supplemented by and later substituted for the oral administration of Rauwolfia and Apresoline

ANEMIC HEART DISEASE

Anemia may affect the circulatory system in two ways Sudden loss of large quantities of blood (more than 20 per cent of the circulating blood volume) results in shock this topic is probably not within the scope of a textbook on pediatric cardiology On the other hand a gradual loss of blood over a long period may cause circulatory alterations by increasing the load on the heart at the time when its efficiency is diminished because of poor coronary circulation

Etiology

The most common causes of anemic heart disease in children today are sickle cell anemia leukemia hypoplastic anemia Mediterranean anemia, anemia of parasitic infection and nutritional or infectious anemia

The degree of anemia required to cause demonstrable evidences of heart disease depends on the duration of the disease and the functional status of the heart It stands to reason that a heart riddled with rheumatic infection will be less able to cope with the extra work imposed on it by the anemia than a healthy heart would be It also seems logical that a moderate anemia of three weeks duration will have fewer circulatory consequences than the same degree of anemia lasting for six months or more With these qualifications it may be stated that serious disturbances probably do not occur with hemoglobin levels higher than 7 0 gm per 100 ml of blood[47]

Physiology

The physiologic consequences of anemia are based on the diminished oxygen capacity of the blood and the consecutive tissue anoxia. This lowered oxygen supply to the tissues by itself and the compensatory mechanisms involved may cause profound alterations in the circulation The following observations have been made on adults with severe anemia by various authors (1) shortened circulation time[44] (2) increase in cardiac output[47] and (3) increase in percentage of oxygen extracted from the arterial blood[1]

Clinical Picture

The clinical picture of anemic heart disease is dominated by the presence of a murmur and moderate or severe cardiac enlargement It should be stated emphatically that there is practically no murmur that could *not* be caused by severe anemia it is totally useless to attempt to determine whether a patient s murmur is organic" or functional" significant" or "insignificant" in the face of severe anemia Evidences of left sided and right sided cardiac enlargement and even pitting edema may be present The electrocardiogram may show changes in the S T segments and T waves over the left ventricle as well as evidences

of mild left ventricular hypertrophy Blood pressure determinations characteristically show a wide pulse pressure with a significant decrease in the diastolic level and an elevation of the systolic level

It should be mentioned here that to the best of my knowledge the changes in the heart in sickle cell anemia are limited to those just described and no increased incidence of rheumatic heart disease exists

Excellent hemodynamic studies of adults with severe anemia exist[47, 93, 310] I am not aware of cardiac catheterization studies on any large series of anemic children

Treatment

The treatment of anemic heart disease consists in slow transfusion with packed cells Extreme caution should be exercised not to correct the severe anemia of these patients too rapidly lest one provoke left ventricular failure and pulmonary edema Small increments of cells administered frequently (probably no more than 5 to 10 ml. per pound at a time every eight to twelve hours) will correct the anemia and abolish the picture of anemic heart disease

CARDIAC TUMORS

Tumors of the heart should be mentioned briefly This subject has been well reviewed recently by Engle and Glenn[108]

Apparently metastatic tumors (bronchogenic carcinoma malignant melanoma and so forth) are more common than primary ones

Among the primary tumors intracardiac myxomas arising in the atria are the most common others worth mentioning are sarcomas and teratomas

The clinical picture in secondary tumors obviously will be dominated by the primary neoplasm Primary tumors may cause symptoms by obstructing the mitral[136] tricuspid or even the aortic orifice[32] They also may cause conduction disturbances congestive failure and progressive cardiac enlargement Significantly enough murmurs are not usually present The electrocardiogram shows progressive evidence of myocardial damage and angiocardiograms may show filling defects Probably cardiac catheterization is not of much use in the diagnosis of cardiac tumors except in the negative sense

The diagnosis almost never can be made with certainty but if it is suspected strongly enough exploratory thoracotomy may be performed

PART THREE

CONGENITAL HEART DISEASE

General Principles

Congenital heart disease is a structural abnormality of the heart probably present from birth There are two qualifying statements in this definition The word structural" excludes cardiac arrhythmias which although they are often present at birth and may give rise to serious cardiac insufficiency once they are abolished may not recur and the patient may be left with a perfectly normal circulation for the rest of his life The statement that these malformations are probably" present from birth seems at first glance unnecessarily vague However since a considerable portion of congenital heart diseases even in this age of excellent pediatric care are not discovered until several weeks or even months after birth there is some question as to when these malformations actually do take definite shape Furthermore as will be brought out later in the discussion of the individual malformations a number of rather common lesions such as patent ductus arteriosus coarctation of the aorta or tetralogy of Fallot, although perhaps recognized as congenital heart disease from the beginning do not assume their pathognomonic characteristics until many months or years later Consequently the question arises whether some of these serious congenital malformations may not in fact develop through the first months or years of extrauterine life from a heart that is only slightly abnormal at birth This point is brought up because I believe that the type of thinking which assumes that the malformation is the same at birth at ten years and at twenty years of age may actually hamper our understanding of the natural history of these anomalies

For purposes of classification and simplification the group of primary myocardial diseases although probably present at birth and certainly representing structural abnormalities has been discussed elsewhere

The lesions to be discussed within the framework of congenital heart disease are (*a*) the abnormal communications between the systemic and pulmonary circuits which include primarily the left to right shunt group (*b*) the valvular and vascular lesions encompassing the right to left shunts together with the obstructive lesions and (*c*) the transposition of great arteries veins or individual cardiac chambers These anatomic entities form the basis of our classification the physiologic variants of each will be discussed separately

Table 18 *Incidence of Major Congenital Cardiac Lesions in Maude Abbott's 1000 Autopsied Cases of Congenital Heart Disease*[1]
(In order of frequency)

Pulmonic stenosis with open ventricular septum	115
Patent ductus arteriosus	105
Coarctation of aorta	85
Ventricular septal defect	62
Complete transposition of the great vessels	49
Pulmonic stenosis with closed ventricular septum	35
Atrial septal defect	33
Dextrocardia	29
Cor biloculare or triloculare	27
Aortic stenosis	23
True truncus arteriosus	21
Tricuspid atresia	16
Aortic atresia	12
Aortopulmonary fenestration	10
Anomalous pulmonary venous drainage	4
Miscellaneous	374
Total	1000

INCIDENCE

The importance of congenital heart disease in pediatric cardiology can scarcely be overemphasized Until the last decade it was believed that rheumatic heart disease was by far the most common form of cardiac disease in children Within recent years however it has become obvious that rheumatic and congenital heart diseases occur with about equal frequency in this age group[183, 81] as a matter of fact in many cardiac centers congenital heart disease is the more common of the two The shift of emphasis toward congenital heart disease in children is due to several factors As has been mentioned before rheumatic fever is on the downgrade Also the spectacular advances in the surgical treatment of congenital heart disease have prompted physicians and parents to seek help for infants with congenital cardiac malformations who twenty years ago would have died at home without any specific diagnosis

No accurate large scale figures are available for comparing the two conditions in the general population However at least ten times as many new patients with congenital heart disease are admitted yearly to the Boston Childrens Medical Center as are admitted with rheumatic heart disease Figures from the Toronto Cardiac Registry[183] indicate that they find more than twice as many children with congenital heart disease as with rheumatic heart disease and that deaths from the former are about seven times as common as from the latter The over all incidence of congenital heart disease is placed at about three per 1000 at birth and one per 1000 at ten years of age according to British authors[34] Richards and her associates studying 60 000 babies in New York placed the over all incidence at 0 83 per cent including stillbirths.[88]

The frequency of the individual lesions is difficult to determine

Table 19 Incidence of Major Congenital Cardiac Lesions in 200 Clinically Studied Cases (Paul Wood[36])

Lesion	Cases
Pulmonic stenosis with ventricular septal defect	40
Atrial septal defect	35
Patent ductus arteriosus	29
Pulmonic stenosis without ventricular septal defect	24
Ventricular septal defect	24
Coarctation of aorta	16
Tricuspid atresia	6
Aortic stenosis	6
Eisenmenger complex	2
Complete transposition of the great vessels	2
Miscellaneous	16
Total	200

Table 20 Incidence of Major Cardiac Lesions in Infants Dying in the First Month of Life (Ober and Moore[60])

Lesion	Cases
Transposition of the great vessels	27
Interventricular septal defect	18
Cor triloculare and cor biloculare	12
Coarctation of aorta	11
Tetralogy of Fallot	4
Miscellaneous	28
Total	100

Table 21 Incidence of the More Common Congenital Lesions in 577 Proved Cases at The Children's Hospital of Boston

Lesion	Cases
Patent ductus arteriosus	101
Atrial septal defect	97
Pulmonic stenosis	83
Ventricular septal defect	68
Tetralogy of Fallot	54
Coarctation of aorta	63
Transposition of the great vessels	36
Eisenmenger syndrome	22
Tricuspid atresia	17
Total anomalous pulmonary venous drainage	13
Truncus arteriosus	11
Single ventricle	4
Ebstein's disease	2
Primary pulmonary vascular obstruction	2
Miscellaneous	4
Total	577

accurately Different figures are obtainable depending on whether one discusses living patients or results of postmortem examinations in addition each author's series varies in terms of age or type of disease Abbott's[1] analysis of 1000 postmortem examinations in respect to the more important congenital heart lesions is presented in Table 18 In

contrast to these data are the clinical results in Paul Wood's[362] series of 200 patients studied by cardiac catheterization (Table 19) In both series pulmonary stenosis and ventricular septal defect are the most common lesions with patent ductus arteriosus occurring next in frequency Table 20 compiled from the autopsy files of the Boston Lying In Hospital [60] shows the frequency of the lesions causing death in the early neonatal period Finally Table 21 summarizes the cases of congenital heart disease at The Children's Medical Center in which the diagnosis was proved either at cardiac catheterization operation or post mortem examination Obviously the vast majority of our patients were children and the interest of our medical and surgical staff is slanted toward the acyanotic group in general

Etiology

The etiology of congenital heart disease is unknown in the vast majority of instances Hereditary factors may play a role since the incidence of congenital heart disease in siblings is significantly higher than in the general population Polani and Campbell in a study of 377 families of children with congenital heart disease found the incidence to be two per 100 families [73] In our experience the same lesions are likely to be repeated in siblings of the same sex we have seen patent ductus arteriosus pulmonary stenosis and tetralogy of Fallot each in two siblings We have also encountered patent ductus arteriosus in a mother and daughter both successfully operated upon Taussig[331] described a family in which the father and four children all had patent ductus arteriosus Walker and Ellis [343] reviewing the literature found forty eight families in which more than one sibling had congenital heart disease In our series of approximately 2500 patients we have fifteen families in which two siblings have congenital heart disease

Other congenital malformations associated with congenital heart disease will be mentioned only briefly Mongolism is commonly associated with persistent atrioventricular canal or ostium primum Coarctation of the aorta is found in conjunction with arachnodactyly and Turner's syndrome Transposition of the abdominal viscera with or without dextrocardia is commonly accompanied by septal defects and transposition of the great vessels Mental retardation and cleft palate with harelip seem to me to be more common in children with congenital heart disease although no statistical confirmation of this clinical impression is available

Environmental influences during pregnancy may be implicated Mothers who contract German measles within the first three months of pregnancy are likely to give birth to children with multiple congenital anomalies including congenital heart disease [3-5] Among these patients patent ductus arteriosus is the most common lesion but atrial septal defect, tetralogy of Fallot and pulmonary stenosis have also been en

countered. Polani and Campbell [3] stated that mothers between the ages of forty and forty four years are more likely to give birth to children with tetralogy of Fallot than are younger mothers. Animal experiments[313, 316] indicate that noxious influences such as anoxia or vitamin deficiency induce pregnant rats to give birth to litters with multiple congenital anomalies including congenital heart disease. In Peru the incidence of cardiac anomalies especially patent ductus arteriosus seems higher in patients living at high altitudes and low oxygen pressures than in those born at sea level.[2] Certain congenital anomalies are more common in one sex than in the other: lesions involving the aorta (coarctation, aortic stenosis) and also transposition of the great vessels seem more common in males whereas atrial septal defect and patent ductus arteriosus are seen more commonly in females. Seasonal influences are significant only in that patients with patent ductus arteriosus are more likely to be born between October and January than at any other time of the year according to Rutstein and his associates.[300] This is due to the prevalence of German measles in the late winter and early spring months.

How these and perhaps many other factors operate in the production of the individual case of congenital heart deformity is impossible to tell in the overwhelming majority of instances. Usually, even after taking the most careful family and prenatal history, the pediatrician cannot give a satisfactory answer as to why the child has congenital heart disease. Further pregnancies should be encouraged, however, especially if the parents are young. The chances that the next child will also have congenital heart disease are only about 2 per cent, as stated previously.

EMBRYOLOGY

The embryology of congenital heart disease will be touched on only briefly. At the second intrauterine week, when the embryo is only 1.5 cm long, the heart begins to take shape. A functional circulatory system has been established by the fourth week; the ventricular septum is fully developed by the eighth week. Consequently, any radical alteration of the architecture of the heart must occur some time between the second and eighth weeks of intrauterine life.

The primitive cardiac tube in the three week embryo is a vertical structure attached dorsally by pericardial folds at its two ends. It consists of four chambers, from caudal to cranial direction: the sinus venosus, the atrial canal, the ventricular canal and the bulbus cordis. This originally straight cardiac tube undergoes changes consisting in torsions, kinkings, development of septums and so forth, and at the same time it maintains the circulation of the embryo.

The sinus venosus, into which the umbilical veins and the vitelline veins drain, is absorbed into the right atrium. The anterior cardinal

veins fuse into an inferior vena cava the posterior cardinal veins contribute to the inferior cava and the coronary venous system

The common atrial canal is divided into left and right atria by a septum primum growing down from the roof of the atrium at the end of the third or the beginning of the fourth week (5 cm embryo) There are two openings in the septum primum—a foramen primum which is in the lowest aspect of the septum posteriorly in the neighborhood of the atrioventricular valve and a foramen secundum which is high in the septum primum approximately where the foramen ovale will be later Under ordinary circumstances the growth of the septum primum and its attachment to the endocardial cushion of the atrioventricular valve completely abolishes the low foramen primum The foramen secundum on the other hand is closed only partially by the growth of a newly formed septum secundum This septum secundum develops about the fifth or sixth week and grows from the posterior wall of the atrium in a crescent shape anteriorly to the right of the septum primum It partially covers the foramen secundum leaving a slitlike opening the foramen ovale As long as the pressure in the right atrium is higher than that in the left atrium this opening allows the flow of oxygenated placental blood from the inferior vena cava into the left atrium

Thus the defects in the atrial septum developing between the fourth and sixth weeks may originate from the foramen primum or secundum A foramen primum defect is low principally posterior and involves a defect of the atrioventricular valves as well In contrast the true atrial septal defect which develops because the foramen secundum was not closed by the septum secundum is usually high dorsal and accompanied by normal mitral and tricuspid valves A patent foramen ovale develops if the septum primum fails to fuse with the posterior edge of the normally developed septum secundum

The common ventricular canal is divided by an intraventricular septum (septum inferius) which starts at the same time as the septum primum of the atria and grows upward and backward toward the common atrioventricular orifice which it divides into the mitral and tricuspid valves This septum is muscular and is an outgrowth of the ventricular wall itself It does not reach the floor of the bulbus cordis and an opening in the ventricular septum is left in the center high up with a downward convex border This opening is closed by a membranous structure growing down partly from the bulbus cordis and partly from the endocardial cushions on the side The ventricular septum as mentioned is usually completely closed by the seventh or eighth intrauterine week

In order for the ventricular septum to divide the common atrioventricular orifice evenly this must shift toward the right failure of this orifice to shift results in tricuspid atresia and an excessive shift gives rise to mitral atresia Failure of the bulbar ridges or the endocardial cushions to form the membranous part of the septum results in most

common forms of ventricular septal defect Defects of the muscular septum are usually ventral and high near the aortic orifice only rarely do they occur in the neighborhood of the apex

The bulbus cordis the most cranial part of the primitive cardiac tube participates in the formation of the aortic and pulmonary orifices the membranous portion of the ventricular septum and the infundibulum of the right ventricle The extracardiac continuation of the bulbus cordis is the truncus arteriosus

The development of the bulbus cordis at the five to seven week period through a spiraling endocardial ridge and its attachment to the divided ventricular cavity is difficult for medical students and cardiologists alike to understand I never really understood it until a cartoon like motion picture demonstrated it very simply I shall not attempt to explain this development of the bulbar part of the heart except to say that failure of the correct bulbar development may give rise to congenital aortic and pulmonary stenosis transposition of the great vessels or ventricular septal defect with overriding aorta If the septum in the truncus communis fails to develop entirely persistent truncus arteriosus results If there is a localized defect in the bulbar ridges aortopulmonary fenestration results

The blood vessels develop at approximately the same time as the heart The principal structures are four large vessels the two ventral and the two dorsal aortas The two ventral aortas grow cephalad from the bulbus cordis parallel with each other In the cervical region they bend backward forming the first branchial artery and grow down both sides of the spine This occurs at the second to third week of intrauterine life Within the next two to three weeks five more connections between the ventral and dorsal aortas develop thus six pairs of branchial arches are formed However all these branches never exist simultaneously their appearance and disappearance are conditioned by the flow of blood and by the time one pair develops another may disappear The first four branchial arches are present at the end of four weeks the next two develop at the end of six weeks The fifth branchial arch disappears quickly in its entirety without normally participating in the formation of any of the extrauterine vessels Eventually the ventral aortas form the ascending portion of the aortic arch and the two external carotids The internal carotids are made up of the third branchial arches and the dorsal aortas The arch of the aorta is the fourth left branchial arch and the descending aorta is the left dorsal aorta The pulmonary arteries are derived from the ventral part of the sixth branchial arch whereas the ductus arteriosus originates from the dorsal part Variations in this normal development form the basis of the many anomalies of the aortic arch

The Fetal Circulation and Its Adjustments after Birth The intrauterine circulation in the human fetus differs from that of extrauterine

life in a number of respects (1) Oxygen is not obtained through the pulmonary circulation but from the placenta through the umbilical vein and the ductus venosus (2) Oxygenated blood does not enter the left side of the heart directly but is emptied into the right atrium through the inferior vena cava From here it is transferred to the left atrium by means of preferential streaming through the foramen ovale (3) The ductus arteriosus is open and carries blood from the pulmonary artery into the aorta This right to left shunt is dependent on the fact that the pulmonary resistance is higher than the systemic resistance in the fetus (4) Because of these various shunts pulmonary and systemic circuits do not receive equal amounts of blood Toward the end of gestation the lungs receive 10 per cent the fetal tissues minus the lungs 35 per cent and the placenta 55 per cent of the total combined ventricular output[94]

The studies of Barcroft[6] and Dawes[94] on newborn lambs and Lind and Wigelius[16] data on human fetuses suggest the following description of the fetal circulation The superior vena cava carries blood about 30 per cent saturated from its usual drainage places into the right atrium Here it is mixed to a certain extent with blood pouring in through the inferior vena cava which is about 65 per cent saturated This inferior vena cava blood represents a mixture of fresh placental blood (80 per cent saturated) and reduced systemic venous blood (25 per cent saturated)

The mixing in the right atrium as indicated is not complete Most of the blood from the inferior vena cava enters the left atrium through the foramen ovale and most of that from the superior vena cava enters the right ventricle through the tricuspid valve The right ventricular output representing a mixture of inferior and superior vena cava blood (50 per cent saturated) enters the pulmonary artery slightly more than half of the pulmonary arterial blood enters the descending aorta through the patent ductus arteriosus and the rest circulates through the lungs and enters the left atrium

Left atrial blood then consists of a mixture of pulmonary arterial blood entering through pulmonary veins (one fifth) and inferior vena cava blood entering through the foramen ovale (four fifths) The oxygen saturation of the mixture in the left ventricle supplying the head and the right arm is about 60 per cent In contrast the left arm and the lower part of the body are supplied by a mixture of left ventricular and patent ductus arteriosus blood with an oxygen saturation of less than 60 per cent As indicated more than half of the combined ventricular output circulates through the placenta via the umbilical arteries to receive oxygen The well saturated umbilical venous blood enters the inferior vena cava through the ductus venosus bypassing to a great extent the liver

What happens at birth to rearrange this complicated setup into the

well known circulatory pattern of extrauterine life? The two principal factors responsible are the major decrease in pulmonary resistance due to the sudden expansion of the lungs and the minor increase in systemic resistance due to the collapse and cutting of the umbilical cord

As the pulmonary resistance drops with the first few breaths blood is diverted from the ductus arteriosus and virtually the total right ventricular output is poured into the left atrium This extra amount of blood in the left atrium and the sudden increase in resistance confronting the left ventricle raise left atrial pressure and result in closure of the flap of the foramen ovale Thus the two circuits are separated the foramen ovale is closed by the increased left atrial pressure and the patent ductus arteriosus is bypassed by virtue of the sudden expansion of the pulmonary arterioles

In this way extrauterine circulation is established within a few minutes or hours That this is however not a complete and irreversible change is demonstrated by the fact that various environmental influences —particularly anoxia—may cause these channels to reopen temporarily this will be discussed in more detail in the section on foramen ovale and patent ductus arteriosus (see pp 284 344)

Physiologic Consequences of Congenital Heart Disease

The clinical picture of individual congenital heart diseases with the physiologic background will be discussed in detail with the individual lesions However certain general principles common to many forms of congenital cardiac malformation may be summarized here

The physiologic consequences of the various congenital cardiac anomalies may be (1) arterial unsaturation that is admixture of unoxygenated blood in the left side of the heart or the aorta (2) overwork in terms of systolic or diastolic overloading of the ventricles and (3) inadequate systemic cardiac output

Arterial Unsaturation Normally the blood in the pulmonary veins the left side of the heart and the systemic arteries is more than 95 per cent saturated with oxygen Consequently if a blood sample obtained from a peripheral artery (femoral brachial and so forth) of the average person is analyzed by the Van Slyke method its oxygen content should be at least 95 per cent of its capacity that is 18 to 19 ml of oxygen per 100 ml of blood Under certain circumstances principally during crying or respiratory insufficiency of one sort or another the oxygen saturation of the peripheral arterial blood may be as low as 92 per cent in apparently healthy people If however the arterial oxygen saturation is 91 per cent or lower arterial unsaturation is present

Cyanosis Arterial unsaturation is not always accompanied by visible cyanosis and cyanosis does not always mean arterial unsaturation The bluish color of cyanosis is the result of an increased amount of reduced hemoglobin in the capillaries Lundsgaard and Van Slyke[5]

Table 22 Oxygen Saturation and Oxyhemoglobin and Reduced Hemoglobin in Normal and Cyanotic Subjects

	Normal			Cyanotic		
	O_2 Sat %	Oxy Hgb gm %	Red Hgb gm %	O_2 Sat %	Oxy Hgb gm %	Red Hgb gm %
Arterial blood	95	14 25	75	75	11 25	3 75
Capillary blood (calculated)	85	12 75	2 25	65	9 75	(5 25)
Venous blood	75	11 25	3 75	55	8 25	6 75

stated that at least 5 0 gm of reduced hemoglobin must be present in the capillaries before clinical cyanosis is noted (Table 22) Since the average person carries 15 0 gm of hemoglobin per 100 ml of blood one third unsaturation would have to exist at the capillary level before 5 gm of reduced hemoglobin would be found Since the capillary blood according to Lundsgaard and Van Slyke normally contains about 2 25 gm of reduced hemoglobin a drop in arterial saturation to 75 per cent will add the additional 3 0 gm of reduced hemoglobin necessary for manifest cyanosis (Table 22) As a matter of fact physicians who frequently see patients with cyanotic disease usually are able to recognize cyanosis in patients whose arterial oxygen saturation is 80 to 85 per cent

One further point to be emphasized is that according to this concept the presence of cyanosis depends not only on the percentage of saturation of the arterial blood but also on the absolute level of circulatory hemoglobin Thus an 80 per cent saturation in a child who has 10 gm of hemoglobin per 100 ml of blood results in 2 gm of reduced hemoglobin in the arterial blood whereas the same percentage of saturation in a person with 15 gm of hemoglobin is accompanied by 3 gm of reduced hemoglobin and thus more definite cyanosis

This then illustrates the point that definite arterial unsaturation may or may not be accompanied by cyanosis depending on the degree of unsaturation and the absolute level of the hemoglobin

On the other hand definite visible cyanosis may be present in patients whose arterial blood is fully saturated These are persons in whom the extraction of oxygen is so complete that reduced hemoglobin is present at the capillary level in amounts greater than 5 gm thus giving the clinical picture of cyanosis with normal arterial oxygen saturation

I am certain that in a book written for pediatricians the fact that "cyanosis" may indeed be imitated by methemoglobinemia or nitrite poisoning needs to be mentioned only briefly Our more experienced older teachers can remember the time when argyria caused by prolonged administration of Argyrol also had to be considered in the differential diagnosis

These two types of cyanosis the central (with arterial unsaturation) and peripheral (without arterial unsaturation) may be distinguished clinically by the fact that the former is much more generalized and severe while the latter is usually milder affecting only the tips of the fingers toes and nose and not the mucous membranes An easy and reliable method of distinguishing between the two types of cyanosis is by direct determination of the arterial oxygen content Peripheral cyanosis may be seen in severe cases of mitral stenosis and pulmonary stenosis without right to left shunt whereas central cyanosis is most likely to occur with intracardiac right to left shunts although it may be caused by pulmonary disease as well

In a patient with central cyanosis arterial unsaturation may occur either because the pulmonary veins themselves do not carry fully oxygenated blood or because the fully saturated blood of the pulmonary veins becomes contaminated by reduced hemoglobin after it has been delivered to the left atrium The first instance is an example of pulmonary cyanosis and can be identified in most (but not all) instances if the inhalation of 100 per cent oxygen raises the arterial oxygen content to 100 per cent or more Patients with pulmonary cyanosis get pink when put in an oxygen tent or when they cry On the other hand if a right to left intracardiac shunt is responsible for the central cyanosis the child will get much bluer when he cries and will not become completely pink in an oxygen tent neither will arterial samples from these patients show a rise to normal oxygen level after inhalation of 100 per cent oxygen The clinical differential diagnosis of cyanosis is schematically presented in Figure 166

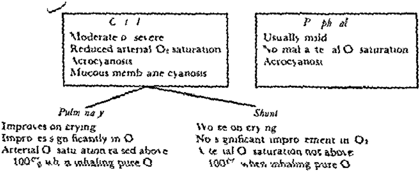


Figure 166 Differential diagnosis of central and peripheral cyanosis

Perhaps a few words could be spent here in explaining the physiology of central cyanosis In the pulmonary type the hemoglobin does not become fully saturated with oxygen in the capillaries of the lung whether the low pulmonary venous oxygen saturation resulting from this is due to the low oxygen tension of the alveolar air or to a barrier between the alveoli and the capillaries it can at least theoretically be corrected by increasing the oxygen tension in the alveolar air Thus ac

counts for the good response to inhalation of 100 per cent oxygen in most of these cases Conversely in instances of shunt cyanosis of the central variety in which the hemoglobin in the lungs does become fully saturated increasing the oxygen tension in the alveolar air results only in a lesser increase in the arterial oxygen content which may be due partly to the increased absorption of oxygen into the plasma and partly to the circulatory adjustments consecutive to oxygen administration

Shunt cyanosis is not as stated in some places *the result of an inadequate pulmonary blood flow it is always due to the addition of venous to arterial blood* For shunt cyanosis to occur two things are necessary (1) a communication between the two circuits such as a septal defect or a patent ductus arteriosus and (2) a resistance to flow of blood on the right side that is larger than the resistance to flow on the left side

The deep bluish color of cyanosis does not have to be illustrated by colored plates it should be familiar to every physician It is most obvious in the mucous membranes of the eyes and mouth but occurs also in the fingers toes and the glans penis Less severe unsaturation may give rise to excessive redness of the lips cheeks and finger tips sometimes the parents of these children remark that this particular youngster has always had a "much better color" than the rest of the family

CLUBBING Under ordinary circumstances then the first result of arterial unsaturation is cyanosis The second slightly later but almost as dramatic consequence is the development of clubbing or hypertrophic osteoarthropathy (Fig 3 p 12) In its fully developed form this consists in a widening and thickening of the ends of the fingers and toes accompanied by convex fingernails Earlier forms of clubbing consist in shininess and tenseness of the terminal phalanges obliterating the wrinkles usually present in this part of the skin This incipient clubbing is usually accompanied by fiery red fingers giving a characteristic picture of early slight arterial unsaturation I have seen full blown clubbing in severely cyanotic children as young as two or three weeks old ordinarily however it does not make its appearance until a child is one or two years old For some reason it appears earliest and most pronounced on the thumbs Physiologic and histologic studies of clubbing indicate that these fingers have an increased number of capillaries with increased blood flow through myriads of arteriovenous aneurysms [42] This is accompanied by an increase of connective tissue in the terminal phalanges

Clubbing in children almost always indicates congenital heart disease of the cyanotic variety although occasionally cirrhosis of the liver subacute bacterial endocarditis lung abscess or even a familial hereditary condition may be responsible for it

POLYCYTHEMIA Polycythemia with increased hemoglobin content is another consequence of arterial unsaturation That a low arterial oxygen content acts as a stimulus to the bone marrow has been amply demonstrated not only on patients with congenital malformation of the

heart but also in people living at high altitudes with low atmospheric oxygen tension[174] The increased oxygen carrying capacity achieved by this method is an extremely useful compensatory mechanism until the polycythemia reaches hematocrit levels of 80 per cent or more At these levels however the benefits derived from the increase in available oxygen are probably outweighed by the disadvantages of the high viscosity The therapeutic implications of these considerations will be discussed later (p 467)

DYSPNEA Dyspnea on exertion is a characteristic feature of patients with arterial unsaturation Exercise demands increased oxygen delivery to the tissues In congenital heart disease with arterial unsaturation this is not possible As a matter of fact in many instances the amount of oxygen delivered may actually decrease on exertion because of the increased right to left shunt resulting from the decreased systemic resistance Under these circumstances dyspnea and hyperpnea occur possibly as a response to the stimulation of chemoreceptors

ANOXIA Anoxic spells consisting in an initial period of uncontrollable crying followed by paroxysms of dyspnea cyanosis unconsciousness and even convulsions occur in certain types of cyanotic congenital heart disease especially in tetralogy of Fallot These alarming episodes are of utmost seriousness they may even lead to the death of the patient Usually they do not follow severe exertion but rather occur early in the morning after a good night's sleep Often they are associated with bowel movements or feeding at other times they cannot be attributed to any single environmental factor Their duration is from a few minutes to several hours In mild cases they may occur once every two or three months whereas in the severest instances a baby may have several of these episodes in one day It is said that these attacks are an expression of cerebral anoxia an unproved hypothesis accepted by most physicians in the absence of a more accurate explanation Anoxic spells seldom make their appearance at birth they usually start at three to four months of age and disappear before the fifth year of life

SQUATTING Squatting described by Taussig[431] is a characteristic posture assumed after exertion by patients with certain types of cyanotic congenital heart disease specifically tetralogy of Fallot and less commonly pulmonary stenosis with an open foramen ovale (Fig 1 p 6) It first makes its appearance at about one and one half years of age or when the child starts walking Social pressure usually though not invariably abolishes it at eight to ten years of age Detailed studies of our own and those of others[6] indicate that oxygen saturation diminished by exercise can be restored to normal more rapidly in a squatting than in a standing position (Fig 167) This may be accomplished either by increasing systemic venous return or possibly by augmenting the peripheral resistance and thus diminishing the degree of right to left shunt

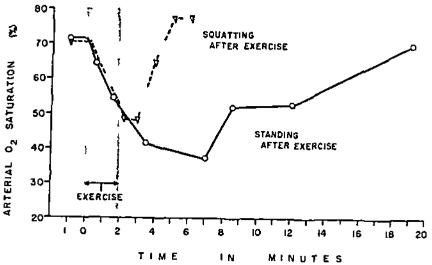


Figure 167 Effect of squatting on exercise induced arterial oxygen unsaturation in a patient with cyanotic congenital heart disease

Overloading of the Ventricles SYSTOLIC OVERLOADING An increase in the resistance to outflow results in systolic overloading of the ventricles In the left ventricle this occurs with aortic stenosis or coarctation of the aorta whereas on the right side it may be the result of pulmonary stenosis or pulmonary vascular obstruction The high pressure in the ventricle is usually best demonstrated clinically by the electrocardiogram Striking changes such as increase in voltage prolongation of ventricular activation time depression of the S T segment and inversion of the T wave allow an almost quantitative estimation of the height of pressure and the degree of hypertrophy in the appropriate ventricle Indirect proof of systolic overloading of the ventricles can also be obtained from the evidences of atrial hypertrophy in the electrocardiogram On the other hand fluoroscopy is almost useless in detecting the presence of a small but concentrically hypertrophied ventricle I have repeatedly encountered patients with maximal aortic stenosis even leading to death in whom the cardiac silhouette was essentially normal This really is not surprising if one bears in mind the few millimeters or centimeters of thickening involved in maximal cardiac hypertrophy

Chronic systolic overloading of the left ventricle is usually well tolerated without symptoms The left ventricle is capable of performing many times its normal work load and thus ensures a normal cardiac output under a wide variety of circumstances This great adaptability has the one disadvantage that it gives no warning to the patient that he is overtaxing his heart hence the cases of sudden death probably from ventricular fibrillation in previously asymptomatic youngsters with aortic stenosis At postmortem examination these patients may show tremendous cardiac hypertrophy with diffuse scarring as a result of ischemia By

the same token congestive failure is a late manifestation of systolic overloading of the left ventricle it occurs in the most advanced cases only and usually at a time when treatment medical or surgical is not likely to restore the effectiveness of the circulation for any length of time

Congestive failure may also be a late manifestation of systolic overloading of the right ventricle in both instances it is a matter of utmost seriousness and often is beyond repair However systolic overloading of the right side of the heart gives warnings sooner than its left sided counterpart If the obstruction is in the lung field the pulmonary arterial hypertension resulting from this obstruction may cause dyspnea on exertion and even at rest possibly through stimulation of local pressure receptors Whether the obstruction is at the pulmonary valvular or at the pulmonary arterial level the right ventricle is frequently unable to exert the maximal effort that the left ventricle is capable of exerting under similar circumstances Thus fatigue due to inadequate cardiac output becomes a prominent symptom Finally a right to left shunt may be established by forcing open a hitherto sealed or small foramen ovale and symptoms referable to arterial unsaturation may appear Thus systolic overloading of the right ventricle in contrast to similar overloading of the left ventricle gives a number of warning signs (dyspnea fatigue cyanosis) before congestive failure sets in

Diastolic Overloading Diastolic overloading of the ventricle in congenital heart disease is the result of an increased volume of blood being offered to it during diastole Patent ductus arteriosus and aortic regurgitation are the commonest causes of diastolic overloading of the left ventricle whereas pulmonary venous anomaly atrial septal defect and ventricular septal defect are the most frequent causes of overloading of the right ventricle The increased volume of blood is handled by dilatation and hypertrophy of the ventricles and an increased stroke output In marked contrast to the situation in systolic overloading the radiologic picture is a much more sensitive index of this type of overwork than is the electrocardiogram Radioscopy reveals a significantly enlarged heart and the pulsations are vigorous under the fluoroscope even in moderate diastolic overloading when the electrocardiogram may still be within relatively normal limits Furthermore striking radiologic evidence in this regard may be obtained in all instances except aortic regurgitation from the presence of pulmonary vascular engorgement A note of caution should be sounded however by pointing out that pulmonary vascular engorgement even with expansile pulsations may not be the result of increased volume but rather of increased pressure in the pulmonary arterial tree

The physical examination is also extremely helpful in detecting diastolic overloading of the ventricles One of the most sensitive indices is the presence of a diastolic rumble at the apex or the lower left sternal border which is common to all hearts with large diastolic volumes on

the left or the right side In free aortic regurgitation it is referred to as the Flint murmur in septal defects it has no special name but has been noted with large left to right shunts at any level My impression is that the presence of a definite shunt diastolic murmur indicates a pulmonary systemic flow ratio of at least 2 1. Another equally valuable sign of diastolic overloading of either ventricle is the presence of a hyperactive cardiac impulse often accompanied by left chest prominence Finally infants with left to right shunts are more likely to have severe respiratory infections even pneumonia

Signs and symptoms of left sided or right sided congestive failure appear much earlier in patients with diastolic overloading but represent therapeutically more approachable problems than the congestive failures resulting from systolic overloading Medical or surgical treatment or a combination of the two may restore practically normal circulatory efficiency to these children with severe congestive failure

Inadequate Cardiac Output Inadequate cardiac output may be the result of a heart failing as a consequence of overloading this is discussed in the section devoted to congestive failure (see p 242)

Another type of maximal limitation of cardiac output without congestive failure is the one in which a congenital anomaly of the heart does not allow a left ventricular output sufficient to maintain life for any length of time Prime examples of this are (1) complete transposition of the pulmonary veins or the great arteries and (2) the complete interruption of the aorta either at the valve or at the level of the arch In these cases death usually ensues within the first few weeks or months of life

Varying degrees of physical underdevelopment are commonly found in patients with slight limitations of systemic cardiac output such as those caused by patent ductus arteriosus or atrial septal defect That this may in some instances be the consequence of the somewhat decreased systemic circulation seems proved by the occasionally spectacular growth and weight spurt experienced by children whose defect is corrected surgically My personal view is however that in many instances the underdevelopment represents either a genetically linked abnormality or is the result of the frequent respiratory infections characteristic of the left to right shunt group rather than the outcome of diminished cardiac output This belief is based principally on three observations (1) Complete operative correction of the malformation seldom exerts a significant influence on the growth curve of a child (2) Large and serious defects may be associated with relatively normal development whereas minor abnormalities may be found in relatively underdeveloped youngsters (3) The infant s growth and development may be increased significantly by the vigorous use of chemotherapy and chemoprophylaxis and the consequent diminution in the number of bouts of pneumonia per year

Although these views concerning the influence of moderately severe

Table 23 Anatomical Classification of the Most Important Types of Congenital Heart Disease

Communication between Systemic and Pulmonary Circuits with Left to Right Shunt
Interatrial communications
Foramen ovale
Foramen secundum
Foramen primum
Interventricular communications
Small ventricular defect (Roger)
Large ventricular defect (? Eisenmenger)
Ventricular defect with pulmonary stenosis
Ventricular defect with aortic regurgitation
Single ventricle
Communications between great vessels
Patent ductus arteriosus
Aortopulmonary fenestrum
Persistent truncus arteriosus
Aneurysm of the sinus of Valsalva
Valvular and Vascular Lesions with a Right-to-Left Shunt or no Shunt at All
Coarctation of aorta
Vascular ring
Aortic stenosis
Aortic regurgitation
Underdeveloped left ventricle with aortic atresia
Mitral stenosis
Pulmonary vascular obstruction syndrome
Pulmonary stenosis
With intact ventricular septum
With ventricular septal defect (tetralogy of Fallot)
Pulmonary regurgitation
Underdeveloped right ventricle with tricuspid atresia
Ebstein's anomaly
The Transpositions
Complete transposition of the great arteries
Incomplete transposition of the pulmonary artery with completely transposed aorta (Taussig Bing)
Incomplete transposition of the aorta with pulmonary artery from the right ventricle (tetralogy of Fallot Eisenmenger complex)
Complete transposition of the pulmonary veins
Incomplete transposition of the pulmonary veins
Transposition of the venae cavae
Dextrocardia
Mirror image with abdominal situs inversus
Isolated
Levocardia with abdominal situs inversus

congenital heart disease on the physical development of children may not be uniformly accepted the fact that the mental development of these youngsters is not influenced by their cardiac output finds universal agreement among workers in this field Patients with severe congenital heart disease cyanotic or not cyanotic may be of normal or inferior intellect

T

LEFT VENTRICULAR HYP

NORMAL ARTERIAL SATURATION (>91%)

PULMONARY VASCULATURE

Diminished ↓	Normal ⟶	Increased ✓ ↑
	Coarctation of aorta	Patent ductus
	Aortic stenosis	Aortopulmonary fenestratic
	Pulmonary stenosis (very mild) without R → L	A – V communis
	Ventricular defect (small)	Ventricular defect (modera
	Primary myocardial disease	

Ta

RIGHT VENTRICUL

NORMAL ARTERIAL SATURATION (>91%)

PULMONARY VASCULATURE

Diminished ↓	Normal ⟶	Increased ✓ ↑
Pulmonary stenosis (severe) without R → L	Pulmonary stenosis (mild) without R → L	Partial pulm vein anom
		Atrial defect
		A – V communis
		Pulmonary hyper tension with } Ventricular defect / Patent ductus

most certainly there are more mentally retarded children among those with congenital heart disease than are seen in the general population That these conditions are probably not causally linked to each other is proved by the fact that there is no direct correlation between the severity of the disease and the intelligence quotient and by the observation that corrective surgery has no influence at all on the mental development of these youngsters

These two points should be stressed so as not to raise the hopes of parents who may expect miraculous improvement in physical or mental growth as a result of corrective surgery for congenital heart disease

24

TROPHY OR DOMINANCE (EKG)

LOW ARTERIAL SATURATION (<91)

PULMONARY VASCULATURE

Diminished ↓	Normal ⟶	Increased ↑
Tricuspid atresia Single ventricle with pulmonary stenosis Ebstein s disease		Truncus arteriosus Single ventricle

25

HYPERTROPHY (EKG)

LOW ARTERIAL SATURATION (<91%)

PULMONARY VASCULATURE

Diminished ↓	Normal ⟶	Increased ↑
Pulmonary stenosis with R → L } Atrial defect, Ventricular defect		Total pulm vein anom
Transposition with pulmonary stenos s		Transposition without pulmonary stenosis
Single ventricle with pulmonary stenosis		Single ventricle without pulmonary stenosis
→ Pulmonary vascular obstruction (? primary)	⟷	Pulmonary vascular obstruction (secondary) } A V communis, Ventricular defect, Patent ductus } with or without coarc tation
		Truncus arteriosus Aortic atresia

Classification

Table 23 presents the classification which will form the framework for the presentation of the individual lesions This schematic division is an at empt to present lesions in a logical sequence without being hin dered by the artificial division of cyanotic or noncyanotic disease It is hoped that this classification will make it easier for the reader to re member all the varieties of congenital heart disease

Tables 24 and 25 present a diagnostic classification of the most im portant forms of congenital heart disease based on the presence of left or right ventricular hypertrophy as shown in the electrocardiogram the presence or absence of cyanosis and the nature of the pulmonary vas culature in the radiogram

CHAPTER 15

Communications between Systemic and Pulmonary Circuits

INTERATRIAL COMMUNICATIONS

Patent Foramen Ovale

The foramen ovale is a slitlike opening in the atrial septum at the site of the foramen secundum of the septum primum As indicated in the discussion on embryology the septum secundum surrounding the foramen secundum in a crescent shape leaves a slitlike opening which is covered in a valvelike manner by the free edge of the septum primum The only important function of the foramen ovale is in intrauterine life when it transmits inferior vena cava blood which has a high oxygen content to the left atrium This is possible because in fetal life the high right atrial pressure keeps the valve of the foramen ovale open Shortly after birth left atrial pressure rises and the flap is pushed tightly against the septum secundum closing the foramen ovale functionally Lind and Wigelius[216] using angiocardiographic methods could not demonstrate a functional opening in the foramen ovale at the end of the first week of life Anatomic patency however may occur for several months and even at the end of the first year of life approximately 50 per cent of all infants have a probe patent foramen ovale Probably complete anatomic closure never occurs in 30 per cent of all persons

√The principal fact to remember about the foramen ovale is that it can allow only a right to left shunt because of the flap which covers it from the left side Consequently any functional communication between the two atria through this opening *must* go from right to left A left to right atrial shunt *cannot* be explained on the basis of a patent foramen ovale Therefore a functional opening of the foramen ovale is possible only in situations in which the resistance to flow in the right side of the heart is higher than in the systemic circuit and therefore right atrial pressure is higher than left atrial pressure (Fig 168) Thus the clinical picture of an open foramen ovale consists in a right to-left atrial shunt in

cases of obstruction at the level of the right ventricular pulmonary valvular or pulmonary vascular level

It should be mentioned however that fairly frequently at cardiac catheterization the left atrium may be entered through a functionally *closed foramen ovale* thus pointing up the fact that passage through the atrial septum by the cardiac catheter does not under all circumstances indicate the presence of an atrial septal defect

In summary the diagnosis of a functionally patent foramen ovale *in extrauterine life should be made only in the presence of a right to left* atrial shunt associated with right ventricular and right atrial hypertension

Atrial Septal Defect Including Foramen Secundum and Foramen Primum Lesions

An atrial septal defect is any opening in the atrial septum not covered by a valve

Incidence

In various authors series atrial septal defect occurs with varying frequency it ranked seventh among Abbott s[1] 1000 cases seen at post mortem examination and second among Wood s[36] 200 and our 577 cases studied by clinical means In approximately 15 per cent of all patients who have congenital heart disease and who live through infancy an atrial septal defect is the principal lesion

Anatomy

The foramen secundum lesion high in the atrial septum may be one large defect or may consist of several small almost cribriform openings It is the result of failure of the septum secundum to completely cover the foramen secundum An associated anomaly of the pulmonary venous drainage is commonly present as well

The foramen primum defect on the other hand is low in the atrial septum is of crescent shape and is the consequence of failure of the septum primum and the endocardial cushions of the atrioventricular valves to fuse It is often associated with deformities of the mitral (cleft anterior cusp) or tricuspid valve or even with a small high ventricular septal defect Because of these associations Edwards and his co authors[107] refer to the foramen primum defect as a persistent atrioventricular canal and discuss it as a more or less complete version of atrioventricularis communis

The size of atrial septal defects varies from at least 1 cm in diameter the smallest clinically significant opening to virtual absence of the atrial septum The average size is about 3 cm

The pulmonary arteries and the cavities of the right atrium and the right ventricle are large and often overshadow the left side of the heart

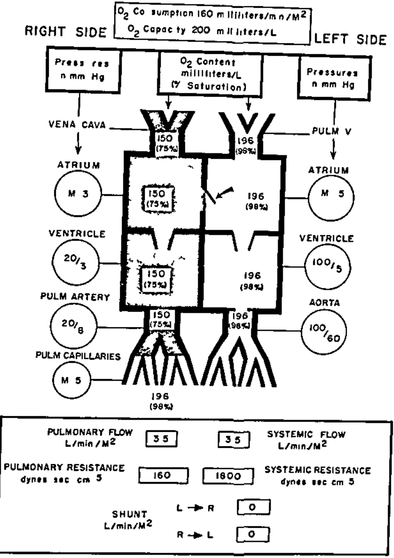


Fig 168 A

and the aorta The wall of the right atrium and the right ventricle is not grossly hypertrophied dilatation dominates the picture Associated left *ventricular enlargement is seen in the foramen primum defects* especially if they are associated with deformities of the mitral valve

PHYSIOLOGY

The flow of blood through an uncomplicated atrial septal defect, ranging from 1 0 to 20 liters per minute per square meter of body surface is always from left to right by virtue of the fact that the pressure in the left atrium is higher than that in the right atrium through the entire cycle This relative hypertension of the left atrium is dependent on the

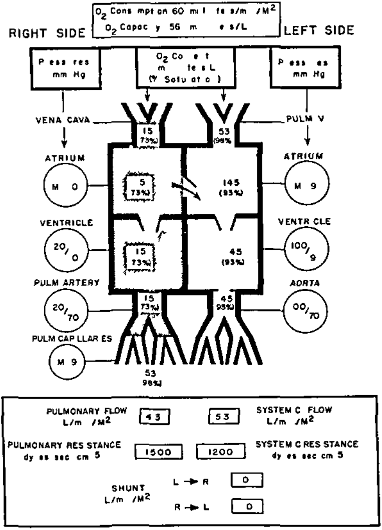


Fig 168 B

F gure 168 D agrammatic presentation of the fact that the flap cover ng the foram n o ale does not perm t the transfer of blood from left to-righ t atrium (*A*) wher as t allows a free flow from r gh t to left if the r gh t atr al pressure is higher (*B*) than the left atrial

difference in the pressure volume characteristics of the two ventricles[100 7 17] Under normal circumstances the left ventricle having to over come the high resistance peripheral circulation is thicker and less dis tensible than the right ventricle working against a low resistance pul monary circu t Thus the left atrium must hypertrophy in order to fill this thick walled chamber and the left atrial pressure curve usually shows a taller A wave and a higher mean pressure than the right atrial

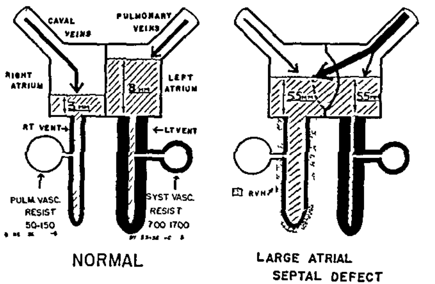


Figure 169 A comparison of the filling of the two ventricles under normal circumstances and in patients with large atrial septal defects Note the low right atrial and high left atrial pressure in the normal person and the equal atrial mean pressure in patients with large atrial septal defects (From unpublished data by W T Goodale and R B Hickler)

pressure curve shows Under ordinary circumstances with an intact atrial septum the left atrial mean pressure is approximately 8 to 10 mm of mercury whereas the right atrial mean pressure is only 4 to 5 mm If a large defect in the atrial septum is created the pressure in the two atria becomes equal with the mean pressure about midway between normal right and left atrial mean pressures At this level of common pressure *in the two atria the right ventricle can be filled easily but the left* ventricle cannot Consequently all the caval blood entering the right atrium and much of the pulmonary venous blood entering the left atrium finds its way to the right ventricle and the pulmonary artery (Fig 169) The flow through this atrial defect probably occurs through the entire *cardiac cycle* Dow[99] *pointed to the teleological argument that the pur*pose of the entire circulation is to ensure an adequate systemic output Consequently if one assumes that a certain percentage of the pulmonary venous blood is shunted from the left to the right atrium then the total pulmonary flow should be sufficient so that the percentage allotted from it to the left ventricle is compatible with life Convincing proof of this hypothesis is afforded by figures which show widely varying pulmonary *blood flows in cases of atrial defect in which systemic indices* are within normal range[49]

The physiologic phenomena just described consisting *in a relatively* normal systemic output with a large left to right shunt and a consecutive increase in pulmonary blood flow are characteristic features of a large foramen secundum defect For practical purposes in these cases no

right to left shunt occurs A complicating factor such as pulmonary vascular obstruction pulmonary stenosis or failure of the right ventricle would have to be present before a right to left shunt through an atrial septal defect of the secundum type could be present

In patients with an ostium primum type of defect which easily blends both anatomically and physiologically into atrioventricularis communis the physiologic situation is somewhat different These patients tend to have an increase in pulmonary vascular resistance similar to that in ventricular septal defects hence right ventricular pressure is more or less elevated and some degree of right to left shunt often occurs simultaneously with the dominant left to right shunt In addition to the increase in pulmonary vascular resistance a further cause of the right to left shunt may be the occurrence of mitral regurgitation which may change the volume pressure characteristics of the left ventricle through dilatation

Two further physiologic variants possible either with a primum or a secundum type of defect should be mentioned briefly One is pulmonary vascular obstruction This does not occur frequently in children (less than 10 per cent of those with atrial defects) When it does occur it is usually associated with a foramen primum type of defect On the other hand pulmonary vascular obstruction in adults with atrial septal defect is relatively common It usually appears in progressive fashion in the late twenties or thirties associated with foramen primum or secundum type of defects and may be the result of the large pulmonary blood flow damaging the pulmonary vasculature through the years

The second variant which should be mentioned is the association of atrial septal defect and a left to right shunt with mild pulmonary stenosis The gradient across the pulmonary valve in these instances is probably due to an unusually large amount of blood attempting to get through a normal pulmonary valve during systole rather than to true anatomic stenosis Additional anatomic reasons may be found in the hypertrophy of the right ventricular outflow tract with the creation of a real obstruction to flow and dilatation of the main pulmonary artery segment

Clinical Picture

The most significant feature in the history of the majority of children with atrial septal defect is the discovery of a murmur Usually this is not noted until the child is well past the neonatal period and in some instances not until the patient is more than five years old

The symptomatology of atrial septal defect in children varies Some patients have no complaints at all whereas in others varying degrees of exercise intolerance episodes of right sided congestive failure and frequent severe respiratory infections are evident (Fig 170) On the whole patients with ostium primum defects are more likely to show symptoms early especially congestive failure whereas lesions of the ostium secun

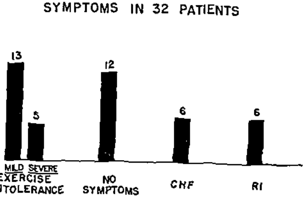


Figure 170 Symptoms in thirty two patients with atrial septal defect[4] CHF = congestive heart failure RI = increased number of respiratory infections

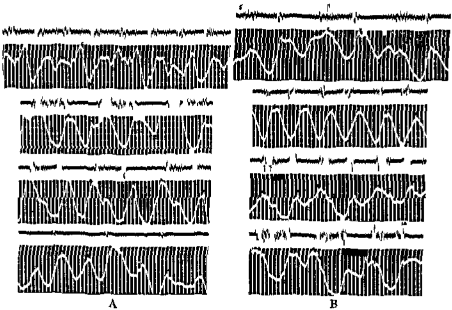


Figure 171 Phonocardiograms correlated with jugular venous pulse in eight cases of atrial septal defect Note the systolic collapse in every instance and the tall V wave[284]

dum cause relatively few symptoms throughout childhood Cyanosis is extremely rare *in the foramen secundum type whereas it is frequently* mentioned in the history of patients with foramen primum defects

On physical examination these patients usually though not invariably appear thin and asthenic *in build The blood pressure is normal* with some tendency toward a narrow pulse pressure The jugular venous pulse shows tall V waves preceded by a systolic collapse (Fig 171) The liver may be palpable but is not tender or pulsating *Left chest deform*ity is common The cardiac impulse is hyperdynamic and heaving It

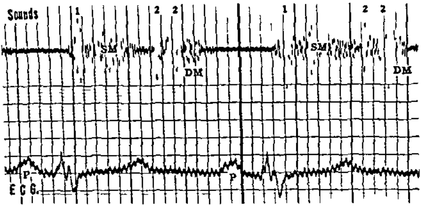


Figure 172 Phonocardiogram of patient with ostium primum type of atrial septal defect

is maximal at the xiphoid process though in foramen primum defect an additional left ventricular type of apical impulse is also palpable A systolic thrill at the second left interspace indicates additional pulmonary stenosis A thrill at the lower left sternal border and apex may occur with foramen primum defects

On auscultation all defects have a systolic murmur In ostium secundum defects this is usually of no more than grade 3 intensity is blowing in character and is heard maximally at the second to third interspace with poor transmission (Fig 109 p 107) The origin of this systolic murmur may be either at the pulmonary valve or at the defect itself In contrast the systolic murmur of a foramen primum defect is much harsher and is heard loudest at the lower left sternal border transmitting well toward the apex and even the back (Fig 172) This murmur near the apex may originate from the mitral regurgitation commonly associated with foramen primum The murmur at the lower left sternal border may be caused by the forceful regurgitant jet through the mitral valve and through the low atrial defect If either a primum or a secundum type of defect is associated with pulmonary stenosis the typical stenotic murmur at the second left interspace will be the dominant feature

Low frequency diastolic murmurs (rumbles) are heard in all atrial septal defects with at least 2 1 pulmonary systemic flow ratio. These either occur immediately beyond the second sound (Fig 111 p 108) or follow the third sound and are best heard just at the lower left sternal border or near the apex An early high frequency diastolic blow may be heard in patients who have an atrial defect associated with pulmonary vascular obstruction and pulmonary regurgitation (Fig 110 p 107)

The first heart sound may be widely split but is of average intensity and is heard best at the apex The second heart sound is always widely split (more than 0 05 second) at the second left interspace and is as a rule of normal intensity A definitely accentuated second sound at the

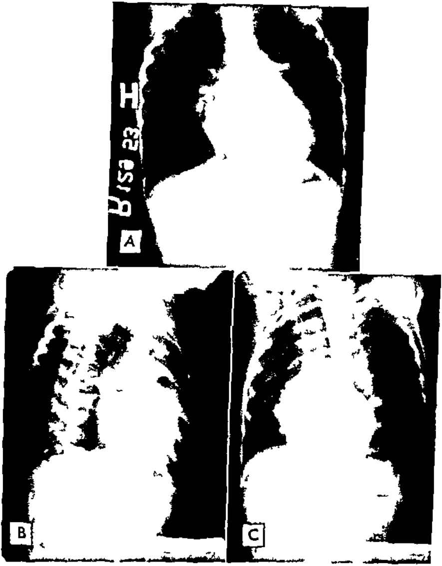


Figure 173 Typical atrial septal defect in posteroanterior left anterior oblique and right anterior oblique views

pulmonary area is heard only in children with pulmonary arterial hyper tension As previously mentioned the third heart sound is heard com monly at the apex

The roentgenologic findings are enlargement of the right side of the heart and enlargement and pulsation of the main stem and the small branches of the pulmonary artery (Fig 173) The total cardiac size is in direct proportion to the size of the left to right shunt (Fig 174) whereas the prominence and pulsations of the pulmonary vasculature do not have direct bearing on the size of the pulmonary blood flow The

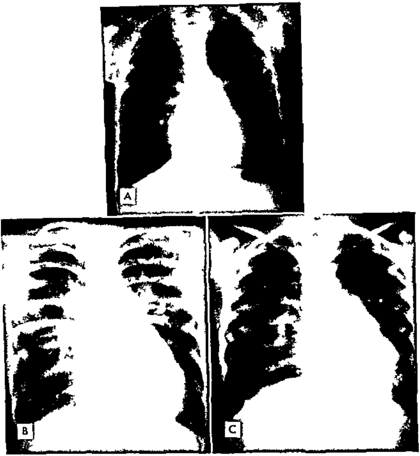


Figure 174 Posteroanterior views of patients with (*A*) small (*B*) medium sized and (*C*) large atrial septal defect

enlargement of the right side of the heart involves the right atrium and the right ventricle to an equal degree No isolated left atrial enlargement is present in uncomplicated atrial septal defect Left ventricular enlargement is suggested radiologically in foramen primum defects with mitral regurgitation and in any patient with congestive failure

The electrocardiogram shows complete or incomplete right bundle branch block in all cases of atrial septal defect (Fig 175) Prolonged P R interval is noted in about 25 per cent of the children who have this anomaly A significant degree of right ventricular hypertrophy with strain (indicating right ventricular hypertension) is an unusual feature of an uncomplicated foramen secundum defect but may be encountered occasionally with a foramen primum lesion Similarly the left ventricular hypertrophy type of chest lead and left axis deviation in the standard limb leads are the rule with foramen primum defects associated with

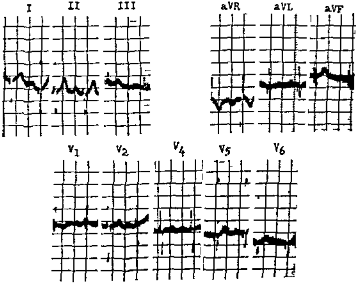


Figure 175 Electrocardiogram of patient with atrial septal defect Note the presence of incomplete right bundle branch block

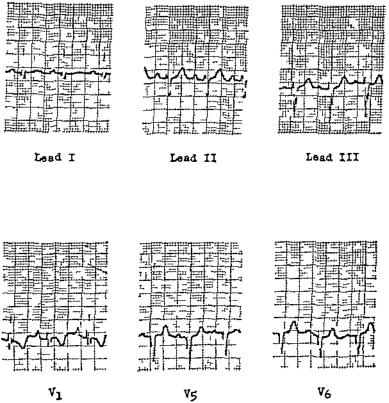


Figure 176 Electrocardiogram of foramen primum type of atrial septal defect Note the presence of left axis deviation in the standard limb leads and right bundle branch block with good left ventricular potentials in the chest leads

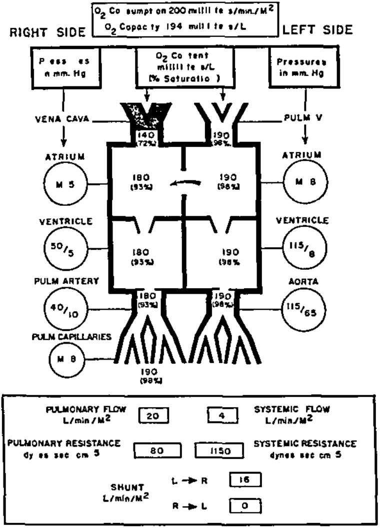


Figure 177 Schematic presentation of cardiac catheterization findings in patient with atrial septal defect (foramen secundum)

mitral regurgitation (Fig 176) whereas they almost never are encoun*tered in ordinary ostium secundum defects*

The findings at cardiac catheterization are as follows (Fig 177)

1 There is an increase in the oxygen content of the right atrium of at least 2 volumes per cent over that of the superior vena cava In foramen secundum defects the appearance of highly oxygenated blood occurs high in the right atrium near the entrance of the vena cava without any *further jump in the ventricle Conversely in a patient with a foramen* primum defect the addition of oxygenated blood is likely to occur near



Figure 178 Radiogram demonstrating the passage of the catheter through an atrial septal defect from right to left atrium

the tricuspid valve and a further increase may occur in the ventricle itself

2 The pressures in the right side of the heart are usually within normal limits in children with ostium secundum defects the chief abnormality being the frequent occurrence of tall V waves in the right atrium Conversely children with ostium primum defects and adults in their twenties or thirties usually have more or less pulmonary arterial hypertension

3 The pulmonary capillary pressure is normal

4 The peripheral arterial blood is usually more than 95 per cent saturated

5 As a result of the large atrial shunt, the pulmonary blood flow becomes many times larger than the systemic flow, the smaller shunts have a pulmonary systemic flow ratio of 2 1 whereas the larger ones may have a ratio of 4 1 or 5 1

6 Occasionally the catheter may pass directly from the right atrium to the left atrium (Fig 178) serving as presumptive evidence of an atrial septal defect Usually this maneuver is easier to execute if the catheter is introduced through the inferior rather than the superior vena cava It should also be reiterated that passage of the catheter through the atrial septum does not by itself indicate the presence of an atrial septal de

fect—it may simply demonstrate an anatomically, but not functionally patent foramen ovale

Venous angiography, *in our experience is not a particularly useful* tool in demonstrating the presence of an atrial defect the dilution of the dye by the tremendous left to right shunt does not permit accurate visualization of the atria

Selective angiography, with the catheter in the right atrium also does not allow a right to left injection of the opaque medium However one of the modifications of the selective technique introduced by the Swedish angiographic team[191] may furnish good visualization of an atrial septal defect In one method the contrast medium is injected directly into the pulmonary artery with initial visualization of the left atrium and secondary opacification of the right atrium through the atrial septal defect In a second method the catheter is passed from the right to the left atrium with injection of the medium directly into the left atrium Finally this same group of investigators using catheters equipped with special inflatable balloons have measured the exact size of the atrial defect by passing the catheter through it and injecting a measured amount of contrast material into the balloon attached to the tip of the catheter After the balloon has been filled to a certain size an attempt is made to withdraw it through the defect The size of the defect is assumed to be identical with the size of the balloon that is just too large to be withdrawn through the atrial defect.

Course and Prognosis

The course of uncomplicated atrial septal defect depends on the size and the location of the opening Apparently small defects are tolerated without any difficulty for many years as a matter of fact they are compatible with normal life expectancy Larger defects with big left to right shunts may cause trouble in two ways. (1) The large diastolic overloading may lead to congestive failure in early adulthood (2) The persistently large pulmonary blood flow may lead to pulmonary vascular obstruction in the late twenties or thirties Once this appears it is usually rapidly progressive and is accompanied by severe exertional dyspnea diminution of the left to right shunt and the appearance of cyanosis

As a rule atrial defects are relatively well tolerated during infancy and childhood Lesions of the ostium secundum almost never cause trouble in children ostium primum defects are more likely to be associated with early congestive failure or pulmonary vascular obstruction

The average life expectancy of patients with atrial septal defects is forty years[49] but some live as long as seventy years

Among the complications of atrial defect rheumatic heart disease should be mentioned first In our series it was present in sixteen of

eighty cases seen at postmortem examination Functionally significant mitral stenosis was present in seven (Lutembacher's syndrome) Sub acute bacterial endocarditis is unusually rare postmortem examination revealed evidence of this disease in only two of the eighty cases Atrial fibrillation does not occur in children with atrial septal defect whereas Wood[363] found it in 10 to 20 per cent of his adult patients

Differential Diagnosis

The following entities should be primarily considered in the differential diagnosis of atrial septal defect

Rheumatic Heart Disease Acquired mitral valve involvement and atrial septal defect may have several features in common In both conditions females are affected more frequently than males the murmur is usually discovered late systolic and diastolic murmurs are commonly heard at the apical or lower left sternal border and the P R interval is prolonged A further complicating factor is the relatively frequent simultaneous occurrence of the two lesions The presence of a basal systolic murmur electrocardiographic evidence of incomplete right bundle branch block expansile pulsations of the hilar vessels on fluoroscopy and absence of isolated left atrial enlargement all point however toward an atrial septal defect Often only cardiac catheterization can distinguish clearly between the two conditions

Ventricular Septal Defect A small ventricular septal defect with a harsh systolic murmur accompanied by a thrill at the lower left sternal border together with electrocardiographic evidence of left ventricular dominance or hypertrophy and radiologic evidence of combined ventricular enlargement and a prominent left atrium can easily be distinguished from an atrial septal defect On the other hand differentiation may be a great deal harder when there is a large ventricular septal defect with pulmonary arterial hypertension (the so called Eisenmenger syndrome) in which the systolic murmur is not so intense and the right ventricle dominates the picture in the electrocardiogram as well as in the roentgenogram In this situation the evidence of pulmonary arterial hypertension expressed in terms of a booming pulmonary closure transient cyanosis and marked right ventricular hypertrophy as shown on the electrocardiogram favors the diagnosis of a large ventricular septal defect

Patent Ductus Arteriosus The typical patent ductus arteriosus with its continuous machinery murmur in which the electrocardiogram shows left ventricular dominance or hypertrophy and roentgenograms reveal left ventricular and left atrial enlargement with a vigorously pulsating large aorta and pulmonary artery is hard to confuse with an atrial septal defect Conversely an "atypical" patent ductus arteriosus without a machinery murmur and with pulmonary hypertension may present a difficult differential diagnostic problem which may be solved only by cardiac catheterization

Transposition of the Pulmonary Veins Complete transposition of the pulmonary veins commonly presents a picture readily distinguishable from atrial septal defect These patients are prone to show congestive failure during infancy and mild cyanosis is the rule If as is commonly the case the veins enter the superior vena cava the radiologic picture is pathognomonic as well

Partial transposition of the pulmonary veins on the other hand may be indistinguishable from an atrial septal defect even with the use of techniques such as cardiac catheterization and dye dilution curves

Treatment

One purpose of medical treatment of atrial septal defect is to bring congestive failure under control The indication for anticongestive treatment is simply the presence of cardiac failure Vigorous treatment along the lines previously indicated often may result in marked and sustained improvement for several years

In addition it is important to try to prevent rheumatic fever in this group of patients In view of the susceptibility of these patients to rheumatic infections their respiratory diseases should be treated promptly and vigorously with antibiotics Many cardiologists believe that continuous chemoprophylaxis should be recommended similar to the regimen for patients with known rheumatic heart disease

The surgical closure of atrial septal defect was first proposed by Murray [49] Other techniques have been successfully worked out by Bailey Gross [155] Swan [41] Brock [54] Bjork [34] and others In the Bailey technique and its modifications the defect is closed by suturing its lower edge to the top of the right atrium In the Gross procedure if the defect is small enough it is sutured directly or through an atrial well the larger ones are closed with a plastic sheet The Swan operation is performed under direct vision in a bloodless field under hypothermia In Brock s technique hypothermia is used only if the defect proves to the exploring finger to be too large to be sutured by one of the simpler techniques In the Bjork procedure the atrial defect is closed by a purse string suture applied to the interatrial groove

No large series is yet available to establish clearly the superiority of one method over another Kirklin[180] summed up the situation by saying that in expert hands any of these methods may be of benefit to some patients Doubtless within the next few years one or two of these methods will prove to be superior and will become the procedure of choice

Mortality figures as presented in the literature are not comparable they vary considerably depending on the technique used the condition of the patient and the anatomic and physiologic situations involved

The best operative risk today is a patient in his early teens or twenties who has a large foramen secundum defect with no complications in this group the mortality rate is approximately 10 per cent

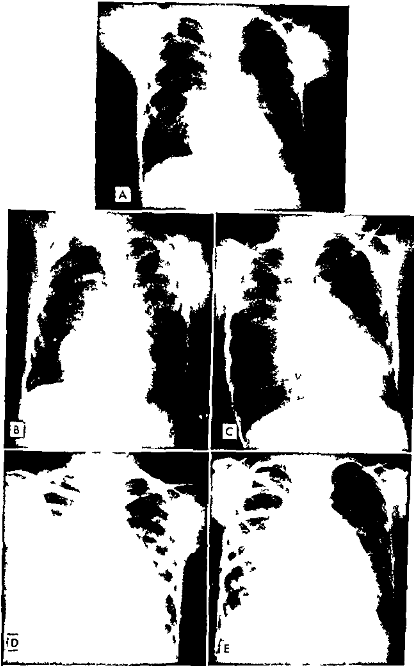


Figure 179 A B C I osteroai tenor and left and right oblique views of patient with ostium primum type of defect before operation *D* Posteroanterior radiogram of the same patient three months po toperative *E* Radiogram of same patient six months postoperative



Figure 180 A and C Preoperative and B and D postoperative radiograms of two patients with closure of atrial septal defect [4]

On the other hand only a few patients with pulmonary vascular obstruction (pulmonary vascular resistance more than one quarter of the systemic resistance) survive surgery they usually die within twenty four to forty eight hours after operation Similarly for patients with foramen primum defects especially those who also have associated mitral valvular disease the mortality rate is approximately 50 per cent Even if these patients survive the operation severe and persistent left sided and right sided failure with large left atria may develop (Fig 179)

On the basis of these considerations patients with an uncomplicated secundum defect and a large (at least 2 1) left to-right shunt should be considered candidates for operation We do not recommend surgery for a patient who has an atrial septal defect with pulmonary vascular obstruction A child with a foramen primum type of defect should not be operated on at the present time either For the time being, patients who have small shunts associated with defects of either the foramen

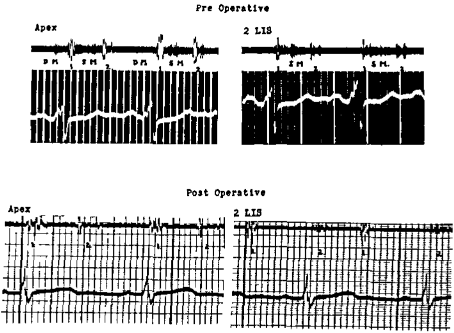


Figure 181 Preoperative and postoperative phonocardiograms in patient with closure of atrial septal defect

primum or secundum with or without pulmonary arterial hypertension are not considered good candidates for surgery

The results of successful surgical closure of an atrial septal defect are gratifying indeed The heart diminishes in size (Fig 180) exercise tolerance increases and congestive failure is no longer present Systolic and diastolic murmurs diminish or even completely disappear (Fig 181) At postoperative cardiac catheterization the pulmonary flow equals or is only slightly larger than the systemic flow However in some cases fluoroscopy reveals a persistent hilar dance and the electrocardiogram may remain unaltered

With regard to preoperative and postoperative care of patients with atrial septal defects a few specific points should be mentioned The oral administration of quinidine sulfate in doses of 100 to 200 mg two hours before operation probably helps to prevent the occurrence of arrhythmias This dosage should be administered every four hours for at least one week postoperatively to avoid the development of atrial flutter or fibrillation During the period when oral administration is not possible the intramuscular route should be used

The postoperative course of these patients may be stormy Left sided and right sided congestive failure should be watched for and the appearance of rales tenderness of the liver and dyspnea are indications for vigorous anticongestive measures including the administration of digitalis and mercury The development of congestive failure in these

✓ *Table 26*

DIFFERENTIAL DIAGNOSIS OF INTERATRIAL COMMUNICATIONS

	CLINICAL PICTURE													PHYSIOLOGY					COURSE & REMARKS
	M				X RA					EKG				SHUNT		PRESSURE			
	S	S	CO	AS	E	W	E				W	L	AA	→	←				
TENT R MEN V LE											0	0			?			N	APA ALLO ONL → HU
FT UM UNDUM	W SPLIT	II III S		LL RUMBL				2	+	+		0	0	+	0	W SLIGHT	4H SLIGHT		SU YMPTOMATIC HROUGH CHILDHOOD D TN EARL OR OUL HOOD OR PVO COM ONL SOC TYP
ST UM IMUM	AC SPLIT	III III L SR @ APEX		LL RUMBL	@	+					+	L				W SLIGHT	SLIGHT +		KEL CAUSE SYMPTOMS IN CHILDHOOD PVO OCCUR FREQUENT SSOCI ION MIT AL INCUS D AL E INVOLVEME

KEY TO TABLES 26 28 29 30 33

P_2	P lmo ary e nd sound	RVH	R ght ve t c la hype t ophy
Syst	Sy tol c	LVH	Left nt cula hype t ophy
Co t	C nt n o s	AH	At al hyp t ophy
D ast	D tol c	BA	B a h al a t ry
AE	At al nla g me t	LLSB	Low left st n l bo de
RVE	R ght ve t cula nla gem nt	LAD	Left x s de at o
LVE	L ft t cula enla gem nt	N	No mal
PA	P lmo ary a t y	CF	Conge t v fa lu
PV	Pulmo ry va cular e	PVO	Pulmona y vas la ob t t o
RB³	R ght b ndle b a h block	TRPV	T a sp s t o of ght p lmo a y ve n

patients may be due to the suddenly increased stroke volume offered to the left ventricle Administration of digitalis may be stopped within one or two months in most instances

Another postoperative complication to be mentioned is transient atrial flutter and fibrillation If prophylactic administration of quinidine did not forestall the onset of this arrhythmia increase in the dosage of quinidine probably will not improve the situation It is our practice to digitalize these patients and keep their ventricular rates below 100 beats per minute within two to four weeks they usually revert to normal sinus rhythm

The urinary output of a patient with an atrial septal defect may be low for twenty four to forty eight hours after operation Maintenance of electrolyte balance guided by frequent determinations of blood chemistry should consist in replacement of the daily water loss by administration of a solution of 5 per cent dextrose and water

The hypotension often encountered in the postoperative period should be combated by appropriate doses of norepinephrine

SUMMARY

Atrial septal defect should be considered in the diagnosis of all patients with a left to right shunt and incomplete right bundle branch block

Auscultatory electrocardiographic and radiologic data may distinguish between high ostium secundum and low ostium primum defects with mitral involvement Cardiac catheterization reveals a significant increase in oxygen content in the right atrium normal right ventricular pressure and large pulmonary blood flow Patients with an uncomplicated ostium secundum lesion with a large shunt should be considered candidates for operation In carefully selected cases the mortality is 10 per cent or less Children with ostium primum defects and those with pulmonary vascular obstruction should not be operated on at present because of the high mortality rates Table 26 presents the salient features in the differential diagnosis of various interatrial communications

INTERVENTRICULAR COMMUNICATIONS

Incidence

On the basis of clinical observations ventricular septal defect was always thought to be the most frequent type of congenital heart disease This is not strictly true Ventricular septal defect was fifth in frequency in Paul Wood s series[367] and fourth in ours If however complications of ventricular septal defect with pulmonary stenosis as well (tetralogy of Fallot and Eisenmenger s disease) are grouped together then it is indeed the most common congenital malformation not only in our series but also in Wood s[382] and Abbott s[1]

Anatomy

The defects of the ventricular septum vary in size from 0 5 to 3 0 cm in diameter

The development of the ventricular septum has already been discussed briefly Most of the defects are probably due to a developmental anomaly of the bulbus cordis rather than of the septum inferius This theory finds good substantiation in the frequent association of these lesions with another defect of the bulbus namely infundibular pulmonary stenosis (see Tetralogy of Fallot)

A great deal of confusion and innumerable classifications often make the nature of ventricular defects hard to understand One of the many obstacles is the difficulty in determining clinically the site of the opening in the ventricular septum and thus identifying the lesion as high or low membranous or muscular in type In fact as was pointed out by Selzer[316] the majority of the defects observed at postmortem examination are high and anterior in the membranous septum connecting the sub aortic portion of the left ventricle with the tricuspid portion of the right ventricle most of these defects include by definition a certain degree of overriding of the aorta In a minority of cases postmortem examination reveals the opening to be low in the muscular septum and in a still smaller number the ventricular septum is entirely absent (single ven

tricle) Probably the anatomic location of the defect and its relation to the aorta are of relatively minor importance The clinical profile and prognosis depend principally on the size of the defect the nature of the right ventricular outflow tract and the condition of the pulmonary vasculature

PHYSIOLOGY

The timing direction and size of a shunt through a ventricular septal defect are determined by well known physical laws the principal governing factors are the size of the defect and the pressure gradient between the two ventricles Because under ordinary circumstances the pressure in the left ventricle is higher through the entire cardiac cycle than it is in the right ventricle an opening in the ventricular septum results in a shunting of blood from left to right principally during systole The height of the systolic pressure in the two ventricles is largely determined by the respective resistances confronting them Minor alterations in the resistances of the two circuits may result in a radical change in the ventricular shunt Similarly a relatively small change in the size of the opening can increase or decrease the flow through the septal defect

In addition to these two main factors the degree of overriding of the aorta also plays a part in determining the dynamics of ventricular septal defect It is debatable how great actually is the role of overriding It seems clear that the greater the degree of overriding (everything else being equal) the greater is the cyanosis At the same time *this factor is probably not as important as was thought in the past and a certain amount of overriding is inherent in any sizable ventricular defect high in the septum*

One final and almost completely unanalyzed factor in determining the dynamics of ventricular septal defect is the effect of the muscular contraction of the septum on the size of the opening during systole

The lesions to be discussed in this section are those with predominantly left to right shunts those associated with predominantly right to left shunts will be discussed in the chapter on valvular and vascular lesions (see p 378)

If the ventricular septal defect is small a considerable systolic gradient with a relatively small shunt may exist between the two ventricles Conversely if the ventricular defect is large no significant gradient between the two ventricles can exist thus the right ventricular pressure will be equal or close to that of the left ventricle Depending on the status of the pulmonary vasculature this right ventricular hypertension may be the result either of a tremendous flow through the ventricular septum or of a marked increase in the resistance in the pulmonary circuit This concept that increased pressure may be the result of large flow increased resistance or a combination of the two is clearly expressed in Ohm s Law—Pressure = Flow × Resistance

Table 27 Classification of Ventricular Septal Defect

Small ventricular septal defect (Roger s disease)	Right ventricular systolic pressure less than left ventricular systolic pressure Small left to-right shunt
Large ventricular septal defect (?Eisenmenger s syndrome)	Right ventricular systolic pressure equal or close to left ventricular systolic pressure with large flow and slightly increased pulmonary resistance or with normal or decreased flow and markedly increased pulmonary resistance
Large ventricular septal defect with pulmonary stenosis	Right ventricular systolic pressure slightly lower than left ventricular systolic pressure Medium gradient across right ventricular outflow tract Moderate or small left to right shunt
Large ventricular septal defect with aortic regurgitation	Right ventricular systolic pressure equal or close to left ventricular systolic pressure Large left to right shunt Wide pulse pressure
Single ventricle	Common ventricular pressure chamber Left to right and right to left shunts in all instances Right ventricular outflow resistance determining clinical direction of shunt

If the ventricular septal defect is associated with mild pulmonary stenosis usually infundibular a moderate degree of left to right shunt may persist in the face of elevation of right ventricular pressure and in association with normal pulmonary arterial pressure

If the ventricular septal defect involves a deformity of the aortic valve as well a combination of ventricular septal defect with aortic regurgitation results

Complete absence of the ventricular septum results functionally as well as anatomically in a single ventricular chamber with a uniform oxygen content in the right ventricle aorta and pulmonary artery and identical pressures in the right ventricle and the aorta

A classification of interventricular communications based on the principles outlined is presented in Table 27

Small Ventricular Septal Defect (Roger s Disease)

Roger [91] in 1879 described two completely asymptomatic patients with identical cardiac findings in one of these postmortem examination revealed a small ventricular septal defect Since that time all asymptomatic small ventricular septal defects have been called Roger s disease Further important contributions to the definition of the syndrome have been made by Taussig [331] Selzer[306] and Wood [36]

With the introduction of cardiac catheterization a new physiologic definition of Roger s disease was sought to supplement the historical one based on anatomy and symptomatology In our clinic Roger s disease is defined as a ventricular defect with a small left to right shunt and without significant right ventricular hypertension " Using Gorlin s orifice formula [14] Wood[36a] calculated the possible size of these defects to be less than 0 5 cm in diameter

The incidence of Roger's disease within the large group of ventricular septal defects is hard to determine with any accuracy since the clinical criteria are unreliable and cardiac catheterization is barely justified in the completely asymptomatic patients Nevertheless it is interesting to note that both in Wood's series and in ours 25 per cent of the ventricular septal defects studied in detail were classified as Roger's disease

Clinical Picture

The outstanding clinical feature is the discovery of a heart murmur usually within the first weeks of life As a matter of fact with only few exceptions the murmur makes its appearance within the first two or three weeks after birth Symptoms are more common than was hitherto believed at least in the group of patients we have had the opportunity to study Mild exercise intolerance fatigue, dyspnea on exertion and repeated severe respiratory infections were noted in at least two thirds of our patients However symptoms usually do not begin until after the second year of age

At physical examination these children are average in development Cyanosis clubbing and redness of the fingers and toes are uniformly absent. A mild degree of left chest prominence is common The cardiac impulse is moderately forceful and usually involves both the region of the xiphoid process and the apex A systolic thrill at the lower left sternal border (but not at the suprasternal notch) is almost always present The blood pressure is normal the jugular venous pulse is not unusual and there is no evidence of congestive failure

On auscultation both the first and second heart sounds are within normal limits A third heart sound is commonly noted at the apex A harsh systolic murmur (grade 3 to 6) (Fig 108 p 106) is present in all patients and is heard maximally at the lower left sternal border particularly over the xiphoid process. This murmur although loud and harsh, usually does not have the grating character and does not transmit as well to the neck and the back as the murmur of semilunar stenosis Transmission to the lower right sternal border may be good

Low frequency, rumbling murmurs are heard at the apex during mid diastole or presystole in about two thirds of the patients (Fig 112 p 108)

Roentgenologic examination reveals the heart to be slightly or moderately enlarged (Fig 182) The enlargement involves mainly the right ventricle with or without simultaneous left ventricular and left atrial involvement The main pulmonary artery is normal or only slightly enlarged and the pulmonary vasculature shows slight engorgement usually without a hilar dance

The electrocardiogram is within normal limits in the vast majority of instances The unipolar chest leads show left ventricular dominance



Figure 182 Roentgenograms in (*A*) posteroanterior and (*B*) left anterior oblique views of patient with small ventricular septal defect

with a slight increase in voltages over the left ventricle Significant degrees of right ventricular hypertrophy and P pulmonale are absent A significant degree of incomplete right bundle branch block is relatively rare The P R interval is usually normal

At cardiac catheterization (Fig 183) the distinguishing features are as follows (1) The oxygen content of the blood in the right ventricle is at least one volume per cent higher than that of the right atrium (2) Right ventricular and pulmonary arterial pressures are normal (3) Pulmonary capillary pressure is normal or only minimally elevated (4) Arterial oxygen saturation is normal (5) The resistance in the pulmonary circuit is usually low (less than 250 dynes per second per centimeter^{-5}) and the pulmonary flow is 1 5 to two times the systemic output (6) Occasionally the catheter enters the left ventricle or the aorta through the defect

Venous angiography is not a particularly useful diagnostic tool in the study of small ventricular septal defects About the only characteristic feature revealed by this type of examination is the poor opacification of the right ventricle in contrast to the sharply outlined right atrium this of course is the result of the dilution of the dye within the right ventricle by left ventricular blood Another and even less reliable angiographic indication is that the right ventricle and pulmonary artery may be seen through several heart cycles while the aorta cannot be visualized clearly at all

Selective angiography with injection of the dye into the pulmonary artery may reveal an interventricular shunt by opacifying the right

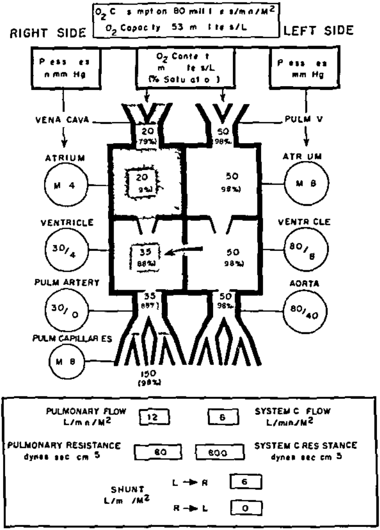


Figure 183 D agram of cardiac catheterization findings in pat ent with small ven tr cular septal defect

ventricle directly from the left heart Differentiation between interatrial and interventricular left to right shunt is not easy under these circum stances

Course and Prognosis

Small ventricular septal defects are thought to be benign lesions Roger s [91] original description implies the innocuousness of this condition Taussig[331] stated that the condition is compatible with long and active life Whether the optimistic view of these and other authors is fully justified is hard to tell since their prognostication is based primarily on

clinical diagnosis and rarely on postmortem findings No accurate long term evaluation of these patients will be possible until some of the children in whom this diagnosis was established early at cardiac catheterization can be followed up through several decades

However from the viewpoint of the pediatrician it can be stated with confidence that at least during childhood these patients do extremely well Although many of them show minimal symptoms these tend not to progress and may even improve We have noted as have others[318] that harsh systolic murmurs (grade 4 to 5 in intensity) may disappear throughout childhood We have assumed as others have that this represented spontaneous closure of a ventricular septal defect As with all other observations of this sort we have no proof of the validity of our assumption

One of the complications of ventricular septal defect is subacute bacterial endocarditis The infective process may affect the margins of the defect or the right ventricular wall opposite the septal opening The incidence of this disease as quoted by various authors ranges from 25 to 51 per cent At present however in my estimation with the widespread use of chemotherapy and chemoprophylaxis considerably less than 15 per cent of the children with ventricular septal defect are likely to contract bacterial endocarditis

Although rheumatic fever is not as common a complication of ventricular defect as it is of atrial defect the two conditions certainly may coexist The sudden development of congestive failure in a patient with hitherto asymptomatic ventricular septal defect ought to raise the suspicion of rheumatic carditis Recognition of this combination is becoming increasingly important since evidence of congestive failure may well be considered an indication for closure of the ventricular septal opening Certainly one s enthusiasm for cardiac surgery would decrease considerably if it could be proved that the patient s symptoms were not due to the presence of the defect but rather were caused by an associated inflammatory lesion

Differential Diagnosis

The differential diagnosis of small ventricular defects may be difficult Several possibilities must be considered

Left to-Right Shunts Defects of the Foramen Primum Atrial septal defects of the foramen primum type are difficult—almost impossible—to distinguish from small ventricular defects on clinical grounds alone Marked incomplete right bundle branch block with left axis deviation usually present with ostium primum may be the most reliable feature in the clinical differential diagnosis Cardiac catheterization may be helpful but often the answer is equivocal At present since operation is not advisable in either condition an accurate distinction between the

two defects is more of an intellectual challenge than a practical necessity

DEFECTS OF THE FORAMEN SECUNDUM A foramen secundum defect is relatively easy to differentiate from a small ventricular septal defect The soft blowing systolic murmur at the second left interspace the absence of a thrill the widely split second sound the exclusively right sided cardiac enlargement and the considerable degree of incomplete right bundle branch block all rather sharply separate foramen secundum defects from the small Roger type of ventricular septal defect Results of cardiac catheterization are usually diagnostic

LARGE VENTRICULAR SEPTAL DEFECTS Large ventricular septal defects can be distinguished from smaller ones by the severity of the symptoms the frequently booming pulmonary closure the relatively softer systolic murmur the suspicion of cyanosis the marked cardiac enlargement involving both ventricles and the pulmonary artery and the right ventricular hypertrophy shown in the electrocardiogram The two entities can be distinguished easily at cardiac catheterization on the basis of the right ventricular pressure level

PATENT DUCTUS ARTERIOSUS If accompanied by the characteristic machinery murmur patent ductus arteriosus cannot be confused with a small ventricular defect The so called atypical patent ductus arteriosus resembles a large ventricular septal defect hence it too should present little difficulty in the differential diagnosis of this lesion

Infundibular Pulmonary Stenosis Infundibular pulmonary stenosis not severe enough to give rise to right ventricular hypertension of more than 60 mm of mercury and thus not producing a right ventricular hypertrophy type of electrocardiogram is extremely difficult to differentiate from a small ventricular septal defect Here again—at least at present—the challenge seems to be intellectual rather than practical since neither condition requires operation and the prognosis for both defects is probably equally good The two conditions have in common a rough systolic murmur and a thrill at the lower left sternal border but the murmur of pulmonary stenosis is somewhat rougher is more diamond shaped and transmits better to the neck and back than the characteristic murmur of a ventricular defect Also in pulmonary stenosis low frequency diastolic murmurs are absent, and the pulmonary second sound is usually diminished However the clinical differential diagnosis is often unsatisfactory and if an accurate diagnosis is desired cardiac catheterization must be resorted to

Infundibular pulmonary stenosis with right ventricular pressure in the systemic range usually though not invariably is associated with tetralogy of Fallot and thus is sharply separated from the acyanotic small ventricular defect However there is a small group of patients who have infundibular stenosis significant right ventricular hypertension and a ventricular septal defect who do not exhibit cyanosis at rest

(acyanotic tetralogy of Fallot) This syndrome still can be distinguished from Roger's disease on the basis of cyanosis on exercise and the right ventricular hypertrophy shown in the electrocardiogram in almost every instance of tetralogy of Fallot

Rheumatic Mitral Regurgitation The systolic murmur of rheumatic mitral regurgitation may resemble rather closely that of a small ventricular defect and both may exhibit apical diastolic murmurs The electrocardiographic and the x ray examinations may be essentially normal in both conditions

A history of rheumatic fever the relatively late discovery of the murmur and its localization to the apex and transmission toward the posterior axilla and the left lung base usually but not invariably suggest the diagnosis of mitral regurgitation even without cardiac catheterization

Mild Congenital Aortic Stenosis Aortic stenosis without significant left ventricular hypertrophy may resemble a small ventricular defect *In both conditions there may be a rough systolic murmur with a thrill* discovered in early infancy a normal sized heart without symptoms and a normal electrocardiogram

The stenotic character of the murmur heard maximally at the aortic area usually makes the differential diagnosis possible even without cardiac catheterization

Subendocardial Sclerosis It may be hard to distinguish subendocardial sclerosis with mitral or aortic valvular involvement or both from a ventricular septal defect as pointed out previously the murmur of both mitral and aortic valvular lesions may closely resemble the one associated with a ventricular defect However in subendocardial sclerosis the heart is diffusely enlarged the pulmonary vasculature is relatively normal and the electrocardiogram may show significant left ventricular hypertrophy Right heart catheterization may reveal no abnormalities or may demonstrate an elevation of right atrial mean right ventricular end diastolic pulmonary arterial mean and pulmonary capillary" mean pressures Some of these patients have complete atrioventricular block they may easily be the group responsible for the legend that complete heart block is common in patients with a small ventricular septal defect

Murmurs at the Lower Left Sternal Border Whether or not harsh murmurs at the lower left sternal border without any other abnormality of the heart revealed clinically or at catheterization are small ventricular septal defects not giving rise to a detectable left to right shunt cannot be decided on the basis of evidence available at present These murmurs may well represent ventricular defects on the other hand they may originate from an entirely normal heart I prefer to say that patients with this syndrome have no significant heart disease

Treatment

No specific medical or safe surgical treatment is available or neces sary at present If a safe surgical procedure can be worked out and if it can be proved by careful follow up studies that patients with Roger s disease suffer long term ill effects from their condition operation should be considered Since at present neither of these two conditions is ful filled surgical correction is not advisable

I recommend that these patients lead a fully active life including sports with the exception of interscholastic competition I prefer to see them once every two or three years at which time they are given an examination including roentgenograms and electrocardiograms I believe that they should receive chemotherapy for all febrile illnesses and chemo prophylaxis for even minor surgical or dental procedures

Summary

The diagnosis of a small ventricular septal defect should be con sidered in patients with congenital heart disease who have slight symp toms and in whom a harsh systolic murmur with a thrill at the lower left sternal border is the only definitely abnormal clinical cardiac finding Cardiac catheterization may provide a definite diagnosis if one is desired Normal findings at right heart catheterization however do not definitely exclude the presence of a small ventricular defect Patients with this condition should be allowed to lead a normal life and should not be considered for surgery unless there are further advances in this field

Large Ventricular Septal Defect (? Eisenmenger s Disease)

Taussig[331] first focused attention on high and large ventricular septal defects in contrast to the small and low ones first described by Roger [91] Selzer[306] emphasized the fact that the size rather than the location of the defect constitutes the principal difference between the two condi tions Other authors particularly Wood[363] and Keith [184] have since de scribed the clinical and physiologic findings in this condition

The incidence of these lesions is probably exaggerated because patients with large defects have more severe symptoms and thus are more likely to seek medical care than are patients with smaller defects Among Wood s sixty cases twenty five probably should be considered to have large ventricular defects In our series of sixty two twenty seven fell into this category Wood using calculations based on Gorlin s formula estimated that the size of the defect in this group ranges be tween 0 5 and 1 5 cm [36] Selzer[306] estimated that the defect in the ven tricular septum in these cases must be at least half the size of the aortic orifice

Clinical Picture

In contrast to Roger s disease patients with a large ventricular septal defect appear sick Usually though not invariably these children

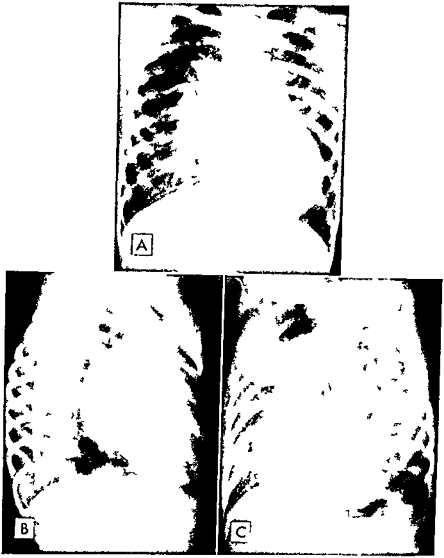


Figure 184. Roentgenograms (posteroanterior and both obliques) of patient with large ventricular septal defect.

have severe symptoms early, sometimes even preceding the discovery of the murmur and the underlying heart disease. All but one of our patients had symptoms by the time they were one year old and more than 20 per cent of them were sick at birth.

The symptoms may be the same as those in Roger's disease but they tend to be more severe. Dyspnea, exercise intolerance, fatigue and repeated attacks of pneumonia are almost invariably present. In addition, episodes of congestive failure and cyanosis occur in about one third of the patients. Hemoptysis, an almost pathognomonic sign of pulmonary

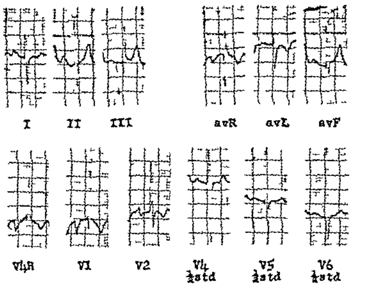


Figure 165 Electrocardiogram of patient with large ventricular septal defect

arterial hypertension, may occur in older children A murmur is usually discovered within the first six months of life

At physical examination these children are poorly developed only rarely does their development place them above the twenty fifth per centile line and most of the time they fall below the third percentile line of a standard developmental chart Cyanosis or redness of the fingers is noted in as many as two thirds of the patients Some chest deformity is almost invariably present A strong, even visible xiphoid impulse often accompanied by an apical thrust is usually observed Pulmonary valve closure is commonly palpable There was no palpable thrill in seventeen of our twenty seven patients this is in marked contrast with cases of small ventricular septal defects in which a thrill is present almost without exception The systemic blood pressure is normal When congestive failure is present the liver is enlarged the neck veins are distended and pulmonary rales are heard The first heart sound is usually accentuated and the second sound is booming at the pulmonary area The first and second sounds are usually narrowly split A third sound may be present at the apex A systolic murmur similar to that mentioned previously is present in all instances but it is usually less intense than in patients with small ventricular defects Twenty two of our twenty seven patients had diastolic murmurs The vast majority of these murmurs were of a rumbling character in mid diastole or presystole in only five patients was there an early insufficiency blow at the second left interspace

Roentgenologic examination reveals considerable cardiac enlargement, involving both ventricles and the left atrium The main pulmonary

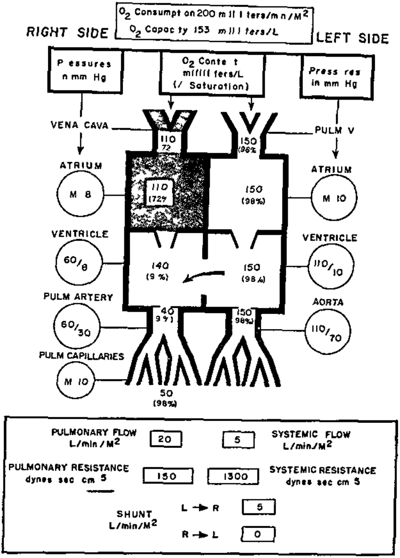


Figure 186 Diagrammatic presentation of findings at cardiac catheterization in patient with large ventricular septal defect

artery segment is prominent, and the pulmonary vasculature is engorged The aorta is of average size (Fig 184) At fluoroscopy the pulsations of the cardiac border are intense the pulmonary artery pulsation is usually stronger than that of the aorta The pulmonary vasculature almost invariably shows intrinsic pulsations

The electrocardiogram shows right ventricular hypertrophy alone or in combination with left ventricular hypertrophy in practically all instances (Fig 185) Right bundle branch block may be present in 25 per cent of the cases Most patients have wide somewhat tall P waves in all standard limb leads suggesting combined atrial hypertrophy Al



Figure 187 Passage of catheter from right ventricle to aorta through a ventricular septal defect

though complete heart block is rare first or second degree atrioventricular block is present in about 10 per cent of the cases

At cardiac catheterization there are significant and severe deviations from normal (Fig 186) The principal abnormalities are as follows (1) The oxygen content of the right ventricle is increased at least one volume per cent over that of the right atrium (2) The systolic pressure is elevated in the right ventricle and the pulmonary artery (3) There is some increase in the pulmonary capillary pressure (4) There may be slight or definite arterial unsaturation (5) There is an increase in the pulmonary vascular resistance with a left to right shunt at least equal to the systemic flow (6) The catheter may enter the left ventricle or the aorta from the right side of the heart (Fig 187)

If the pulmonary resistance is higher than the systemic resistance the shunt through the ventricular defect is entirely right to left and no significant increase in oxygen at the right ventricular level occurs These patients will be discussed with those having pulmonary vascular obstruction even though in certain respects they belong in this group of ventricular septal defects

If the pulmonary resistance though significantly higher than the top normal of 250 dynes per second per centimeter^{-5} is lower than or equal to the systemic resistance left to right or bidirectional shunting occurs through a large high ventricular septal defect The left to right

shunt may vary from 1.5 to 10 liters per square meter per minute The right to left shunt if present amounts to between 1 and 4 liters per square meter per minute when the patient is at rest All in all the anatomic differences between the heart of a patient with pulmonary vascular obstruction and a ventricular septal defect the heart of another with Eisenmenger's syndrome and that of a third with a high ventricular septal defect may be slight indeed The principal difference is in the status of the pulmonary vasculature the role of overriding of the aorta although real is probably of secondary importance Eisenmenger's syndrome is nothing but a large ventricular septal defect with a significant and progressive increase in pulmonary resistance

Angiocardiography may be somewhat more revealing in this group of patients with right to left shunts than in those with Roger's disease Nevertheless so far neither venous nor selective angiocardiography has proved to be the most valuable tool in the study of these patients

Course and Prognosis

As pointed out previously patients who have large ventricular septal defects are likely to have severe symptoms and may even die from infection or congestive failure in early infancy No accurate figures on the mortality rate during the first year of life are available It is my impression however that among all the patients with communications between the systemic and pulmonary circuits these are the ones most likely to terminate early

If with the help of nature antibiotics and anticongestive measures these babies survive the first year or two a remarkable change may occur They are likely to start growing and gaining in weight they become less subject to pneumonia evidences of congestive failure disappear and their exercise tolerance improves It is amazing to see a child with a very large heart severe chest deformity a loud murmur and an abnormal electrocardiogram perform practically normally for many years Paralleling these welcome changes there may be a little more cyanosis the evidences of right ventricular hypertrophy in the electrocardiogram may progress and the pulmonary vasculature may not seem as engorged as it had been earlier

Eventually however these patients start going downhill again severe exercise intolerance develops hemoptysis occurs and increasing cyanosis appears Death in congestive failure ensues when they are in their early twenties or thirties probably with the diagnosis of Eisenmenger's syndrome

Obviously some temporary adaptation takes place in early childhood In my opinion—substantiated by some physiologic and pathologic data—the precarious period of early infancy is due to the large left to right shunt through the ventricular defect As a consequence the systemic output is low right ventricular output is tremendous and pulmonary

arterial "pulmonary capillary" and left atrial pressures are high These physiologic phenomena may well explain poor growth congestive failure pulmonary edema and pulmonary infections During the period of adaptation—the plateau of late childhood and early adolescence—the cross sectional area of the pulmonary vasculature decreases by the development of medial and later intimal changes in the arterioles[9] This elevates the pulmonary vascular resistance diminishes the left to right shunt and makes the development of pulmonary edema less feasible through the now thickened vessel walls

After a number of years the changes in the pulmonary vasculature progress again the flow of blood through the ventricular defect is more in the right to left direction and finally death results from right sided congestive failure and inadequate oxygen uptake

A second and probably less common adaptive mechanism is one in which the increased resistance to right ventricular outflow occurs at the infundibular level rather than in the lung field This gives rise to the syndrome of ventricular septal defect with pulmonary stenosis (see p 322) A diminution of the left to right shunt with all its beneficial effects can be achieved in this fashion without the danger of progressive pulmonary vascular disease inherent in the first type of compensation If the infundibular pulmonary stenosis caused by the development of thickened muscle bands becomes progressively more severe this syndrome may lead to so called acyanotic and later even typical tetralogy of Fallot

A third but most unlikely cause for improvement in a patient with a large ventricular septal defect is the closure of the opening itself Theoretically this is possible it is postulated in the literature repeatedly but we have never seen it proved

The complication of ventricular septal defects by associated bacterial endocarditis or rheumatic carditis has already been discussed (see Roger s disease)

Differential Diagnosis

The problems of differential diagnosis in cases of large ventricular defect concern primarily the other defects in which there is a left to right shunt

Foramen Primum A defect of the foramen primum especially if it involves a small portion of the ventricular septum as well may be mistaken for a large ventricular septal defect The most important distinguishing feature is the absence of severe right ventricular hypertension and its clinical consequences (loud pulmonary second sound and marked right ventricular hypertrophy as shown on the electrocardiogram) in the foramen primum lesion The differences are almost always revealed by cardiac catheterization

Foramen Secundum A lesion of the foramen secundum is very different from a large ventricular septal defect In addition to the dis

tinctive features enumerated in the discussion of Roger s disease there is the added distinction that an uncomplicated defect of the foramen secundum almost never is accompanied by significant pulmonary arterial hypertension in children

Small Ventricular Septal Defect See page 306

Patent Ductus Arteriosus Atypical patent ductus arteriosus or aortopulmonary fenestration cannot with certainty be distinguished by clinical means from a large ventricular septal defect The more evidence of left ventricular hypertrophy there is in the roentgenogram and the electrocardiogram the wider the pulse pressure and the higher at the left sternal border the murmur is located the more a patent ductus arteriosus should be suspected The similarities are so striking that I believe for practical purposes a patent ductus arteriosus should be considered seriously in the differential diagnosis of every severely sick child with a ventricular septal defect Cardiac catheterization and even exploration should be contemplated in every such case

Single Ventricle A single ventricle is only quantitatively different from a large ventricular septal defect the symptoms and signs are more severe At catheterization the increase in the oxygen content of the right ventricle is larger and the prognosis is poorer

Truncus Arteriosus Truncus arteriosus with large pulmonary arteries may be considered in the differential diagnosis Ordinarily the characteristic shape on the roentgenogram the presence of a continuous murmur and the usually more severe course make the distinction pos sible on clinical grounds alone Occasionally the fact that the second sound is completely single may also draw attention to this anomaly Angiocardiography usually establishes the diagnosis

TREATMENT

Successful treatment of large ventricular septal defects is perhaps the greatest challenge remaining in the field of congenital heart disease

The pediatrician and the cardiologist can save these babies lives by using chemotherapy chemoprophylaxis and anticongestive measures intelligently and liberally

Two surgical approaches to the problem have been proposed One a palliative method designed by Muller and Dammann [18] aims at the creation of some pulmonary stenosis by constricting the main pulmonary artery from the outside thus diminishing the left to right shunt and protecting the pulmonary vasculature This is an attempt to duplicate one of the natural adaptive mechanisms mentioned earlier (p 319) It is an ingenious method and may well be the procedure of choice until closure of the defect becomes a safe and easy procedure The main objection to this palliative approach is that it may easily result in a degree of stenosis comparable to that seen in tetralogy of Fallot Also the results of the procedure are not strikingly different from some of the

spontaneous improvements seen through the years in patients with large ventricular defects. Finally, permanent closure of the defects seems so close at hand that one hesitates to recommend a palliative operation for which the mortality rate is about 20 per cent. None of these objections is really prohibitive, however, and I believe that if one encounters a large ventricular defect with considerable pulmonary flow in a child who is extremely sick, the Dammann Muller[48] procedure certainly ought to be considered.

Direct closure of the ventricular defect has been attempted by a number of surgeons. Murray,[19] Mustard,[200] Lillehei[14] and recently Kirklin[104] have all successfully operated on patients with ventricular septal defects. The principle underlying all attempts so far is that of creating a bloodless operative field by using a pump oxygenator. Lillehei[214] and his group used one of the parents in a dramatic cross circulation experiment. Mustard and Keith[200] used a simple pump and monkey lungs for oxygenators. Kirklin and his associates[104] used a Gibbon type of pump oxygenator. None of these methods has been perfected; all of them could be considered, however, in an emergency. Certainly some method will be worked out within the next few years for efficient closure of ventricular defects.

Once this technical obstacle is overcome, however, an almost equally difficult problem—the indications for closure of ventricular defects—will remain. It must be clearly emphasized that many of the difficulties of patients with ventricular septal defects are only partly due to the defect proper and the shunt associated with it. A considerable portion of the trouble is probably due to the presence of pulmonary vascular obstruction. What effect closure of the defect will have on a patient with pulmonary vascular obstruction cannot be stated with certainty, but if the experience gained from patients with patent ductus arteriosus or atrial septal defect and pulmonary vascular disease can be applied to patients with ventricular defects, they will still have difficulties. Although technically the solution to the problem of ventricular defects may be close at hand, the selection of patients for operation may still be difficult for a number of years.

Summary

Patients with large ventricular septal defects are seriously ill. Symptoms of congestive failure, underdevelopment and severe respiratory infections usually begin in infancy. The diagnosis is based on the presence of a rough systolic murmur at the lower left sternal border, accompanied by clinical, electrocardiographic and roentgenologic evidences of pulmonary arterial hypertension. If a child survives the first two or three years, he usually gets better and may lead a relatively normal life for one to two decades. Cardiac catheterization is indicated to provide an understanding of the physiologic mechanism and to differentiate these

patients from those with atypical patent ductus arteriosus As soon as adequate surgery is available and the indications for surgery have been worked out operation should be considered

Ventricular Septal Defect with Mild Pulmonary Stenosis

Ventricular septal defect with pulmonary stenosis depending on the relation between systemic and pulmonary resistance includes a number of clinical entities ranging all the way from the exclusively left to right shunt group[50, 94, 98] to the tetralogy of Fallot The condition to be discussed here is one in which the shunt through the ventricular defect is exclusively or predominantly left to right because of the relative mildness of the pulmonary stenosis the other entities will be discussed in the section on Pulmonary Stenosis

Incidence

Ventricular septal defect with pulmonary stenosis is a relatively common defect there were thirteen such instances in our sixty two cases of ventricular defect Wood[365] found eleven in his series of sixty patients

Anatomy

The majority of these defects are high in the ventricular septum Their size varies greatly but probably most of them are large (0 5 to 1 5 cm)

Physiology

The degree of pulmonary stenosis may be relatively slight It is not always clearly demonstrable anatomically but it is always present in a physiologic sense a significant pressure gradient (more than 15 mm of mercury) between the right ventricle and pulmonary artery is always found at cardiac catheterization In spite of the increase in resistance at the right ventricular outflow tract the total systemic resistance is still higher than the total pulmonary resistance thus the left to right shunt through the ventricular septal defect

Clinical Picture

Perhaps the clinical picture resembles that of Rogers disease more than that of a large ventricular septal defect A murmur was the only complaint in a third of our patients the others had various degrees of dyspnea and fatigue and had frequent respiratory infections Cyanosis is a relatively common presenting symptom in this group but not as common as in children with large ventricular septal defects

The murmur is usually discovered early in all our cases it was present before the child was one year old and often it was noted at birth The age at which other symptoms begin varies from birth to eight years

At physical examination many of these children are well developed and *proportionately nourished* *More than half of the patients in our* group ranked at or above the twenty fifth percentile line on our standard development charts Frank cyanosis is rare Left chest deformity is the rule with few exceptions The cardiac impulse is maximal at the xiphoid process with an occasional left ventricular type of impulse as well A pronounced thrill is always present at the left sternal border in most cases this is maximal at the lower left sternal border but in some instances it appears to be stronger at the upper left sternal border Rales and hepatomegaly are noted in the few patients who have congestive failure Blood pressure is usually normal

At auscultation the heart sounds are usually not remarkable although the second sound at the pulmonary area is sometimes diminished in intensity and splitting A harsh stenotic systolic murmur is noted at the midsternal or lower left sternal border in every instance Although in many cases it is impossible to distinguish this "stenotic" murmur from the murmur of a ventricular defect without pulmonary stenosis differentiation may be attempted on the basis of criteria enumerated previously (p 311) Occasionally one has the distinct impression that a ventricular septal defect type of systolic murmur heard over the xiphoid process can be differentiated from a stenotic murmur noted at the mid left sternal border such a fine differentiation between two systolic murmurs of similar character occurring simultaneously may be impressive to an audience of medical students but is of little practical value and is full of pitfalls Low frequency early or mid diastolic murmurs are present at the lower left sternal border and the apex in about half of the cases early blowing diastolic murmurs are rare and always indicate pulmonary arterial hypertension even in the face of pulmonary stenosis

Roentgenologic examination reveals cardiac *enlargement involving* both ventricles (Fig 188) The main pulmonary artery is usually normal or diminished in size and pulmonary vascular engorgement without hilar dance is present In at least 25 per cent of our cases there was a right aortic arch Occasionally the left atrium seems prominent in the right anterior oblique view

The electrocardiogram showed right ventricular hypertrophy or *combined ventricular hypertrophy in ten of our thirteen cases Left* ventricular dominance was present in three of the thirteen In two of these the right ventricular pressures were less than 50 mm of mercury in the third the right ventricular pressure was 100 mm but the ventricular defect opened beyond the point of maximal stenosis Incomplete right bundle branch block is commonly observed There is usually no significant P pulmonale

Cardiac catheterization (Fig 189) reveals the following data (1) A left to right shunt is present at the ventricular level (2) There is a pres



Figure 168 A Posteroanterior (B) left anterior oblique and (C) right anterior oblique radiograms of a patient with ventricular septal defect and pulmonary stenosis with a left-to-right shunt

sure gradient between the right ventricle and the pulmonary artery (3) The "pulmonary capillary" and brachial arterial pressures are normal (4) There is normal or borderline low arterial oxygen saturation (5) The pulmonary flow is appreciably larger than the systemic flow and pulmonary resistance (valvular and vascular) is always a fraction of the systemic resistance (one sixth to one twenty fifth)

The right ventricular pressure ranges from less than 50 mm of mercury up to systemic levels A gradient of 50 mm of mercury or more implying anatomic stenosis was present in only three of our thirteen patients in the others the gradient varied from 15 to 40 mm The stenosis always seems to be infundibular occasionally the ventricular defect opens in the infundibular chamber beyond the point of maximal stenosis In

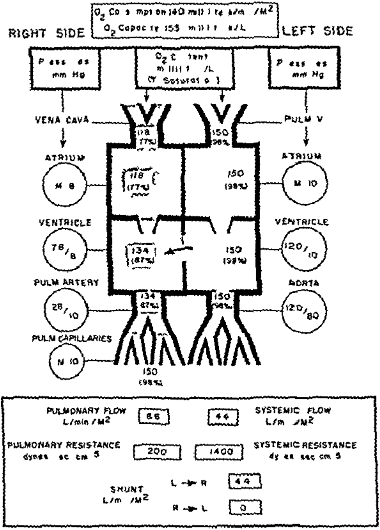


Figure 169 Schematic representation of cardiac catheterization findings in patient with ventricular defect and pulmonary stenosis

our cases the size of the left to right shunt varied from 2 to 15 5 liters per minute per square meter An additional right to left shunt was demonstrated at the time of catheterization in only one patient However *circumstances which bring about a shift in the relationship of systemic to pulmonary resistance* (such as crying or exercise) *may cause right to left shunts in this group of patients* A ratio of systemic to pulmonary resistance as high as 1 2 was noted in the one patient who had simultaneous left to right and right to left shunts

Selective angiocardiography with injection into the right ventricle may be helpful in the diagnosis of these conditions by demonstrating the

nature of the stenosis and perhaps revealing the site of the right to left shunt However more information can usually be obtained by cardiac catheterization

Course and Prognosis

The course of a patient with ventricular septal defect and pulmonary stenosis is a fascinating one Most of these patients have little trouble during infancy and early childhood in this respect they resemble patients who have Roger s disease A few of them however are severely ill and have signs and symptoms identical to those seen in children with large ventricular defects

As time goes on usually by the time they are four to six years of age evidences of left to right shunt diminish pulmonary vascular engorgement becomes less definite and the apical diastolic rumble disappears Coincidentally pulmonary stenosis increases sometimes to the point at which the picture becomes similar to that of tetralogy of Fallot At other times a salutary balance is established between the left to right shunt and the pulmonary stenosis resulting in complete freedom from symptoms and normal pulmonary vasculature

The incidence of complicating subacute bacterial endocarditis and rheumatic fever is about the same as in straightforward ventricular septal defect

Differential Diagnosis

The differential diagnosis of ventricular septal defect with pulmonary stenosis is essentially the differential diagnosis of its component parts (see appropriate paragraphs in the sections on small and large ventricular septal defects and pulmonary stenosis) However special emphasis should be placed on the following entities

Small Ventricular Septal Defect Sometimes it is impossible to differentiate a small ventricular septal defect from a ventricular defect with pulmonary stenosis without resorting to cardiac catheterization The distinguishing points have already been discussed (see p 311)

Large Ventricular Septal Defect Large ventricular defects should not present any real problem in the differential diagnosis because of the evidences of pulmonary arterial hypertension commonly inherent in this lesion

Infundibular Pulmonary Stenosis Infundibular pulmonary stenosis with definite diminution of the pulmonary vasculature and absence of an apical diastolic rumble is usually not hard to distinguish from the combination of ventricular defect and pulmonary stenosis

Patent Ductus Arteriosus Seldom is ventricular septal defect with pulmonary stenosis confused with typical patent ductus arteriosus However an atypical patent ductus arteriosus may sometimes be thought of in patients in whom the ventricular defect and the left to right shunt

occur beyond the point of maximal stenosis at the infundibular level The reason for this is that in these instances the right ventricle is excluded from the flow work inherent in the ventricular defect and the left ventricle carries the entire shunt work load much as it does in patent ductus arteriosus therefore left ventricular dominance or even left ventricular hypertrophy may be manifest in the electrocardiogram in spite of moderate but definite pulmonary stenosis

Treatment

In certain fortunate instances treatment may not be necessary at all the two lesions may act on each other in a compensatory fashion

However if the left to right shunt through the ventricular defect is large as manifested by significant cardiac enlargement and pulmonary vascular engorgement complete repair of the situation should be contemplated when a suitable operation is perfected

If the effects of pulmonary stenosis overshadow the ventricular defect (marked exercise intolerance cyanosis squatting) a shunt operation may give temporary relief This will be discussed in the section on Tetralogy of Fallot Direct relief of the pulmonary stenosis without simultaneous closure of the ventricular defect should not be undertaken lest it cause a significant increase in the left to right shunt

Medical management of these patients includes allowing them full range of activities using chemotherapy and chemoprophylaxis as indicated and treating manifestations of congestive failure if they occur

Summary

Ventricular septal defect with pulmonary stenosis should be suspected from the clinical features characteristic of both lesions—a stenotic systolic murmur accompanied by an apical diastolic rumble pulmonary vascular engorgement and right ventricular hypertrophy as revealed by the electrocardiogram Cardiac catheterization establishes the diagnosis Indications for repair of the ventricular septal defect and for shunt operation are discussed elsewhere Relief of pulmonary stenosis alone without repair of the ventricular defect is contraindicated

Ventricular Septal Defect with Aortic Regurgitation

Ventricular septal defect with aortic regurgitation represents a distinct clinical syndrome which gains significance by its close resemblance to a large patent ductus arteriosus

Incidence

This anomaly was probably first mentioned by Laubry and Pezzi in 1921 [0] More recently Wood and his associates[36] reviewed the literature briefly and discussed the two cases which they encountered within the group of sixty ventricular septal defects studied At The Children's Med

ical Center in Boston we also have had two patients with this condition among our sixty two patients with ventricular defects In addition within the past five years at least five more such patients have been operated on at our hospital with the presumptive diagnosis of patent ductus arteriosus

Anatomy

Anatomically the lesion consists in a large defect in the membranous ventricular septum, associated with distortion and incompetence of one or more of the aortic cusps

Physiology

The load is carried almost entirely by the left ventricle and the left atrium The left ventricular stroke volume must provide for the needs of the peripheral circulation the regurgitant volume into the left ventricle through the aortic valve and the left to right shunt In addition to the increased left ventricular volume work inherent in the lesion increased pressure work is also often required by the elevation of the systemic systolic pressure The ventricular septal defect may result in elevation of the right ventricular pressure work, as well

Clinical Picture

Clinically this lesion resembles a large patent ductus arteriosus A murmur is usually discovered in the neonatal period Symptoms of dyspnea fatigue and frequent severe respiratory infections manifest themselves early. On physical examination these patients are usually stunted and frail considerable left chest deformity is present and the cardiac impulse is maximal at the apex and rocking A thrill is palpable along the entire left sternal border Evidences of right sided and left sided failure are common The pulse pressure is wide with diastolic readings close to zero

At auscultation one is impressed with the loud (grade 5 to 6) systolic and diastolic murmurs audible along the entire precordium By careful analysis and phonocardiography the murmurs can usually be separated into a grade 6 systolic component maximal at the xiphoid process and a grade 4 to 5 systolic diastolic usually to and fro but sometimes continuous murmur at the mid and upper left sternal border These murmurs as expected from their maximal intensity transmit well throughout the precordium neck and back In addition apical low frequency mid diastolic and presystolic murmurs also can be heard The first heart sound is moderately accentuated and the pulmonary second sound is loud if it can be distinguished at all from the extremely loud systolic diastolic murmur (Fig 190)

On roentgenologic examination the cardiac enlargement is maximal and involves both the left and the right ventricles though the former

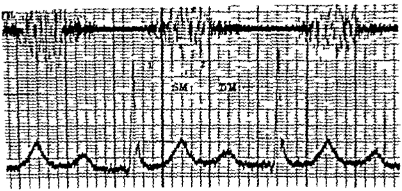


Figure 190 Phonocardiogram of patient with ventricular septal defect and aortic regurgitation.



Figure 191 Posteroanterior and oblique views of patient with ventricular septal defect and aortic regurgitation

usually predominates (Fig 191) Left atrial enlargement is the rule The main pulmonary artery and the aorta are both prominent and the pulmonary vasculature is engorged and shows expansile pulsations

The electrocardiogram reveals marked left ventricular hypertrophy with strain and little if any evidence of right ventricular hypertrophy (Fig 192)

Cardiac catheterization (Fig 193) reveals the following data (1) A large left to-right shunt is present principally at the ventricular level with an occasional slight increase in oxygen content at the pulmonary arterial level as well (2) There is moderate right ventricular and pulmonary arterial hypertension (3) There is slight or moderate elevation of the "pulmonary capillary" pressure with wide systemic blood pressure and rapid brachial artery upstroke time (4) Arterial oxygen saturation is normal (5) The pulmonary flow is commonly three to four times the systemic flow pulmonary resistance may be slightly elevated, and sys

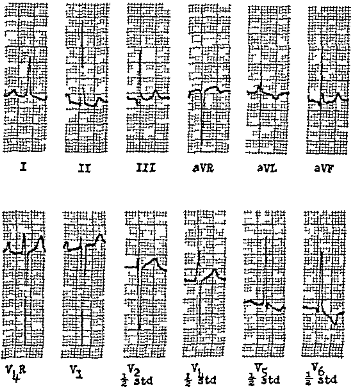


Figure 192 Electrocardiogram of patient with ventricular septal defect and aortic regurgitation Note the marked left ventricular hypertrophy

temic resistance is normal (6) The catheter may pass from the right ventricle to the left ventricle and the aorta (Fig 194)

Angiocardiograms have not been performed on either of our patients but retrograde aortograms were done in both Significantly these did not show opacification of a ductus arteriosus however in one of our two patients the aneurysmal dilatation of the regurgitant aortic cusps was demonstrated in the aortogram (Fig 195)

Course and Prognosis

These patients usually have increasing disability with development of congestive failure and death occurs before the end of the second decade One of our patients had bacterial endocarditis from which he recovered uneventfully

Differential Diagnosis

Large patent ductus arteriosus is about the only important differential diagnostic problem to be considered The similarities are striking both are essentially large left to right shunts with systolic and early diastolic murmurs wide pulse pressure and left ventricular hypertrophy The murmur in the ventricular defect with aortic regurgitation is maxi

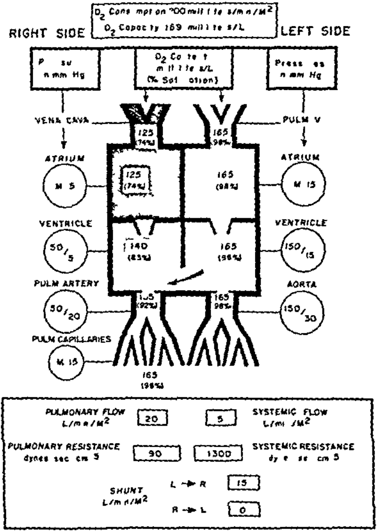


Figure 193 Diagrammatic presentation of findings at cardiac catheterization in patient with ventricular defect and aortic regurgitation

mal at the lower left sternal border and is likely to be to-and fro in character (Fig 132) whereas the murmur of patent ductus arteriosus is usually heard best at the upper left sternal border and is continuous (Fig 113) The pulmonary vascular engorgement is usually not as pronounced in ventricular defect with aortic regurgitation as one would expect from a patient with patent ductus arteriosus of comparable size None of these clinical hints is infallible however Even cardiac catheterization may not provide a definite answer to the question since an increase in oxygen content at the ventricular level may be seen with large patent ductus arterio-



Figure 194 Passage of catheter from right ventricle to aorta through a ventricular septal defect.



Figure 195 Retrograde aortogram in patient with ventricular defect and aortic regurgitation Note the aneurysmal dilatation of the sinus of Valsalva

sus and pulmonary regurgitation although the main increase in patent ductus arteriosus is at the pulmonary arterial level Retrograde aortography may be the best method short of exploratory thoracotomy for differentiating the two conditions The present practice at our institution is to catheterize all these patients and if the answer is equivocal to proceed with thoracotomy as the one means of getting a definite answer as to whether or not a patent ductus arteriosus exists This policy obviously is not universally recommended Its applicability depends to a great extent on the estimated safety of exploratory operations versus the dangers of retrograde aortography

Treatment

No surgical treatment is available for these patients at present The medical management should be conducted according to the principles outlined in the section on large ventricular septal defects

Summary

Ventricular septal defect with aortic regurgitation is similar to large patent ductus arteriosus The to and fro character of the murmur coupled with its localization at the lower left sternal border should make the cardiologist suspect this lesion instead of a large ductus Cardiac catheterization or even thoracotomy may be necessary to confirm the diagnosis

Single Ventricle

From the physiologic viewpoint the complete absence of a ventricular septum should not be regarded as a situation basically different from a large ventricular septal defect Often after a thorough study using all the available clinical and laboratory methods the question of whether one is dealing with a single ventricle or a large ventricular defect becomes one of semantics alone

Because of the rather distinctive anatomic situation and the relatively poor prognosis however it may be worth while to discuss this anomaly under a separate heading but within the group of defects of the ventricular septum

Incidence

This is a rare anomaly It was present in only ten of our 577 cases of proved congenital heart disease Abbott reported thirteen in her series of 1000 cases observed at postmortem examination

Anatomy

The typical case is distinguished by relatively normal atrioventricular valves opening into a single ventricular chamber separated from this common ventricle by a muscular ridge is a rudimentary outflow

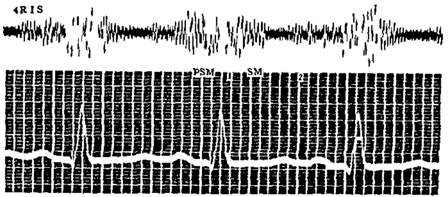


Figure 196 Phonocardiogram of patient with single ventricle Note the loud presystolic and systolic murmur at the fourth right interspace

chamber the remainder of the bulbus cordis giving rise to one or both great vessels In most of the cases reported the aorta arises anteriorly from the small chamber and the pulmonary artery originates posteriorly from the main ventricle constituting some degree of transposition of the great arteries Instances in which the pulmonary artery or both vessels originate from the rudimentary outflow tract have also been reported clinically as well as pathologically

PHYSIOLOGY

The effects of a single ventricle on the circulation depend to a large extent on the relative resistances to outflow in the pulmonary and systemic circuits If the pulmonary outflow tract or the pulmonary vascular bed offers a high resistance to flow the prevailing shunt is from right to left and the patient is cyanotic Conversely if the pulmonary outflow tract and the vascular bed are wide open with low resistance the shunt is almost exclusively in a left to right direction and there is no trace of cyanosis Obviously all gradations between these two extremes exist

CLINICAL PICTURE

The clinical picture is extremely variable depending on the anatomic and physiologic situation Most frequently the presenting complaints are poor development dyspnea and fatigue with or without cyanosis Heart disease is usually discovered at birth or shortly thereafter

At physical examination most of these children are grossly underdeveloped In one of our patients however development was within the fiftieth percentile range Although deep cyanosis is apparent in some of these patients several weeks of hospital observation may fail to reveal any trace of cyanosis in others Left chest deformity is common Dextrocardia may be present A thrill was palpable in half of our patients in two of them it was felt maximally at the right sternal border and

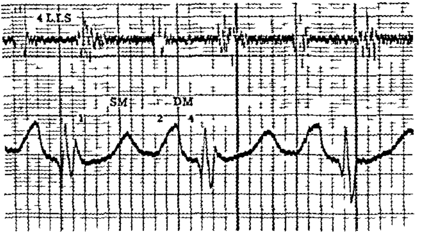


Figure 197 Phonocardiogram of patient with single ventricle Note the faint systolic and diastolic murmur at the fourth left interspace

in others at the left sternal border and apex The blood pressure is usually within normal limits Left sided and right sided congestive failure is common The intensity and splitting of the heart sounds are variable Loud systolic murmurs at the left and right sternal borders often stenotic in character and originating presumably from the gradient between the common ventricle and the outflow tract, are heard in almost every instance Only one of our patients did not have a well defined systolic murmur Loud mid diastolic and presystolic murmurs sometimes at most unusual places (such as the lower right sternal border) are heard relatively frequently (Figs 196 and 197)

The electrocardiogram may reveal a variety of patterns and is characteristically bizarre Theoretically the presence of a single pattern in all the unipolar chest leads would be expected, and this is found in a few patients In others left ventricular hypertrophy is demonstrated Finally straightforward right ventricular hypertrophy may be encountered (Fig 198)

Roentgenologic and fluoroscopic examinations may show considerable pulmonary vascular engorgement (in patients in whom there is free communication of the pulmonary artery with the ventricular cavity) (Fig 199) or ischemic lung fields (in patients with pulmonary obstruction) (Fig 200) Cardiac enlargement may be moderate or marked The pulmonary arterial segment is usually convex because of the superimposition of the rudimentary chamber The heart often is in the right side of the chest

Cardiac catheterization (Fig 201) reveals the following data (1) There is a large increase in the oxygen content of the ventricle (3 0 to 4 volumes per cent) over that of the right atrium the oxygen content is the same in the ventricle systemic artery and pulmonary artery

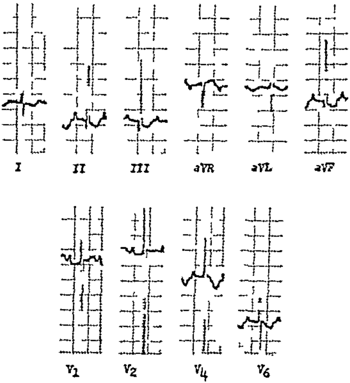


Fig 198 A

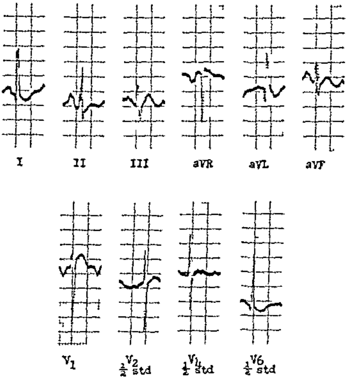


Fig 198 B

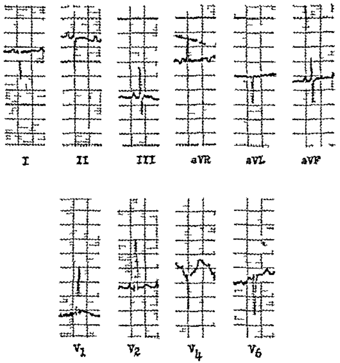


Fig 198 C

Figure 198 Electrocardiograms of patients with single ventricle *A* Note the suggestion of left ventricular hypertrophy in the standard and unipolar limb leads and the single pattern rS in the four chest leads *B* Left ventricular hypertrophy *C* Right ventricular hypertrophy

(2) The systolic pressure in the ventricle is identical to that in the systemic artery the pressure in the pulmonary artery is sometimes diminished because of associated pulmonary stenosis (3) The "pulmonary capillary" pressure is normal or slightly increased and the brachial arterial tracing is normal (4) Definite arterial unsaturation is present ranging from 91 to 60 per cent (5) A dominantly left to right or right to-left shunt exists depending on the respective resistances as discussed earlier (6) The catheter swings around the entire ventricular cavity without showing any change in pressure or oxygen content

Angiocardiography in the left anterior oblique view is helpful in demonstrating a simultaneous and homogeneous filling of the entire ventricle

Course and Prognosis

The outlook for a patient with a single ventricle is as variable as the clinical and anatomic picture The patient with low pulmonary resistance may "bleed" into the lungs may not be able to maintain an adequate systemic output, and may die within the first few days of life

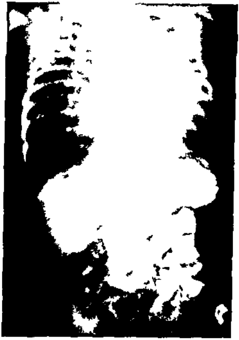


Figure 199 Posteroanterior radiogram of patient with single ventricle and pulmonary plethora

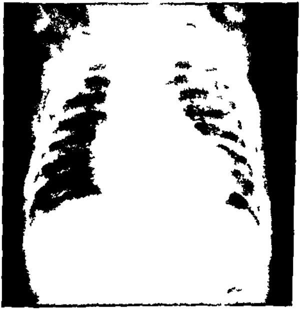


Figure 200 Posteroanterior radiogram of patient with single ventricle and pulmonary ischemia

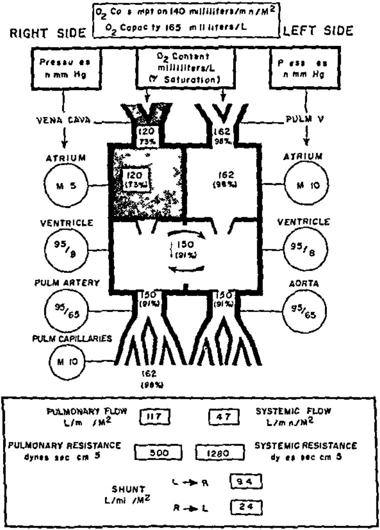


Figure 201 Diagrammatic presentation of cardiac catheterization data in patient with single ventricle

Others with similar but less severe lesions may live for a few weeks or months but finally die without cyanosis in congestive failure and pneumonia Conversely patients with single ventricle and maximal pulmonary stenosis may die early with severe cyanotic disease and evidences of anoxia The patients who fare best are those who have some degree of pulmonary stenosis—enough to allow good systemic and pulmonary flow these children may live for several years with practically no symptoms At present we are following up three such patients in our clinic two of them age seven and one age nine According to Abbott [1] the average age of these children at death is approximately seven years

DIFFERENTIAL DIAGNOSIS

The conditions to be thought of in the differential diagnosis of single ventricle encompass almost the entire range of congenital heart disease since depending on the physiologic setup both the left to right and right to left shunt groups must be considered

Cyanotic Group TETRALOGY OF FALLOT Tetralogy of Fallot may be similar to a single ventricle with right ventricular outflow obstruction this is easily understandable since the only essential difference between the two conditions is the size of the ventricular septal defect Nevertheless clinical identification of single ventricle is frequently possible on the basis of the presence of diastolic murmurs a convex pulmonary arterial segment and an atypical electrocardiogram (frequently showing left ventricular hypertrophy)

TRICUSPID ATRESIA Tricuspid atresia may commonly be associated with single ventricle It can be distinguished electrocardiographically by the presence of clear cut and marked left ventricular hypertrophy with significant P pulmonale

TRANSPOSITION OF THE GREAT ARTERIES In transposition of the great arteries there is marked cyanosis with pulmonary vascular engorgement and a very large heart

Left to-Right Shunt Group VENTRICULAR SEPTAL DEFECT Ventricular septal defect with or without pulmonary stenosis differs principally from single ventricle in the matter of degree and accurate differentiation even with cardiac catheterization is frequently impossible As a rule the patient with a ventricular septal defect is not as sick as the patient with single ventricle

PATENT DUCTUS ARTERIOSUS Atypical patent ductus arteriosus may be considered in the differential diagnosis of single ventricle with left ventricular hypertrophy as shown on the electrocardiogram Cardiac catheterization is the only accurate means of distinguishing between these two conditions

TREATMENT

No method has yet been designed by which the absent ventricular septum can be replaced whether a prosthesis will be developed for this purpose is questionable since its attachment in the face of complete absence of a septum might be impossible The procedure advocated by Muller and Dammann [48] of creating pulmonary stenosis of a mild degree in patients with a single ventricle and a large pulmonary blood flow may have some use in a palliative sense

A shunt procedure—Potts or Blalock's—for the relief of cyanosis is indicated in cases of single ventricle with severe pulmonary stenosis and has been successfully performed in our hospital as well as in other institutions Although on the basis of present day physiologic principles there is no reason why the anastomotic procedure should not be perfectly

Table 28

DIFFERENTIAL DIAGNOSIS OF INTERVENTRICULAR COMMUNICATIONS

	CLINICAL PICTURE													PHYSIOLOGY					COURSE & REMARKS
	C	M	CONT DIAST		X-RAY	PV	VE		PV		E	RV		SHUNT →	→L	PRESSURE		[illegible]	
SMALL VSD (ROGER'S)		[illegible]				+			—					SLIGHT		OR SLIGHT	OR SLIGHT		BENIGN COURSE SLIGHT IF ANY SYMPTOMS
LARGE VSD (EISENMENGER'S)		[illegible]		APE															STORMY INFANCY IF SURVIVING GOOD CHILDHOOD DEATH 20-30s OF OR PH
VSD & PS		[illegible]		[illegible]	OR							+					OR		[illegible]
VSD & AR		[illegible]		[illegible]															STORMY COURSE & EPISODES PNEUMONIA, DEATH IN ADOLESCENCE
SINGLE VENTRICLE		[illegible]		[illegible]	OR														[illegible] SURVIVE TO [illegible] TEENS

See Table 26 (p 303) for explanation of symbols

suited for symptomatic relief in these cases the results are not wholly satisfactory the mortality rate seems higher and the results in the survivors although sometimes good on the whole are not as favorable as they are in tetralogy of Fallot Nevertheless if a patient with a single ventricle has pulmonary stenosis with severe cyanosis and limited exercise tolerance a shunt procedure is indicated However the relatively unfavorable outlook should be made clear to the parents before operation

Medical treatment in terms of anticongestive measures and control of infections should be utilized when indicated

SUMMARY

Single ventricle may present itself clinically either as a somewhat atypical tetralogy of Fallot or as a large ventricular defect In the former instance radiologic electrocardiographic and auscultatory findings may establish the diagnosis In the latter case only cardiac catheterization can furnish proof by demonstrating that the oxygen content and systolic pressure are the same in the ventricle as they are in the systemic artery The prognosis is poor Surgical relief by means of a shunt procedure is available for patients in the cyanotic group

COMMUNICATIONS BETWEEN GREAT ARTERIES

A communication between the great arteries in extrauterine life may be either the result of persistence of a normal fetal structure (such as patent ductus arteriosus) or the outcome of a profound developmental anomaly (such as aortopulmonary fenestration or truncus arteriosus) These defects are totally different in terms of statistical significance

as well as in their influence on cardiodynamics they are grouped together for discussion only to preserve the anatomical framework of the presentation

Patent Ductus Arteriosus

Incidence

Statistically patent ductus arteriosus is a very important lesion In Abbott s series it was the second most common deformity in Woods it occupied third place and in our own series it was the most common lesion (We have classified ventricular defect and tetralogy of Fallot separately)

Anatomy

The ductus arteriosus much like the foramen ovale is a structure necessary for maintenance of fetal circulation it is through this channel that blood reaches the descending aorta from the pulmonary arterial system The ductus connects the pulmonary artery at its bifurcation with the aorta just beyond the origin of the left subclavian artery Embryologically it originates from the dorsal part of the sixth branchial arch Patten [67] computed the cross section of the ductus at term to be about half that of the aorta

Physiology

In the fetus because of the relative hypertension in the pulmonary circuit the ductus carries blood from the pulmonary artery to the aorta In the congenital anomaly to be discussed here the ductus arteriosus not only remains patent beyond the normal time limits but is also considerably larger and thinner than the expected normal at birth Whether the large size of the ductus is secondary to the fact that it has not closed at the normal time or whether the size is the deciding factor in its patency cannot be answered with certainty at present How much of the right ventricular output actually is diverted through the ductus into the descending aorta at various stages of fetal life is a matter of lively controversy It is likely however that in the last weeks of fetal existence more and more blood goes through the lungs and enters the pulmonary vascular system thus reducing materially the flow through the ductus Barcroft and his associates demonstrated the flow of blood through the ductus of a lamb for about five minutes after delivery after that "functional closure occurred [6] Dawes also studying lambs found that closure started within ten to fifteen minutes after birth but he observed flow through a reduced lumen with increased velocity and accompanied by a thrill for several hours or days [94]

In man using angiocardiography Lind [15] demonstrated anatomic patency for several months and Hultgren[173] proved the presence of

functional patency for several days after birth Christie[30] on the basis of postmortem examinations demonstrated anatomic patency in 12 per cent of babies two months old and in 1 per cent of those one year old

On the basis of these data it seems increasingly clear that the closure of the ductus is not an instantaneous affair but rather something that begins during the last weeks of intrauterine life and is not completed for several months after birth

What are the factors which influence normal closure of the ductus arteriosus? There is no one answer to this important question Kennedy[138] demonstrated conclusively that in guinea pigs at least increased oxygenation of the fetal blood causes a muscular constriction of the ductus Land[15] stated that interference with respiration in newborn infants causes a functionally closed ductus to reopen Hultgren and his associates found evidence to suggest that breathing low oxygen mixtures may reopen the ductus in babies less than three days old[1 3] In addition it seems logical to assume that pulmonary vascular changes and pulmonary arterial pressure have an important effect on closure of the duct al though there are no conclusive studies proving this point Finally that factors other than those present at birth may profoundly affect the fate of the ductus arteriosus is proved by the observation that if the mother has German measles during the first trimester a patent ductus arteriosus commonly occurs in the baby The familial occurrence of the anomaly has also been described in the literature I have seen siblings and a mother and daughter with patent ductus arteriosus

The hemodynamic changes occurring at birth specifically the lowering of the pulmonary arterial resistance due to respirations and the increase in systemic resistance due to the occlusion of the umbilical cord result in a reversal of the pressure relationship between the aorta and the pulmonary artery Thus a gradient exists across a patent ductus from the aorta to the pulmonary artery *resulting in a flow of blood from the systemic to the pulmonary circuit* Under these circumstances the left atrium the left ventricle and the pulmonary artery handle an appreciably larger volume of blood than do the right atrium and the right ventricle In most instances the extra amount of blood may be accommodated within the pulmonary arterial bed without a significant increase in pressure Because of the runoff from the aorta into the pulmonary artery aortic diastolic pressure is low and systemic pulse pressure widens

The hemodynamic considerations thus outlined are typical of the vast majority—probably 90 per cent—of the cases of patent ductus arteriosus However in a small number of instances encompassing the largest patent ductus pulmonary arterial hypertension with pulmonary vascular obstruction is present This situation as will be demonstrated later changes the entire clinical and hemodynamic picture In addition to or even supplanting the left ventricular flow work pulmonary arterial

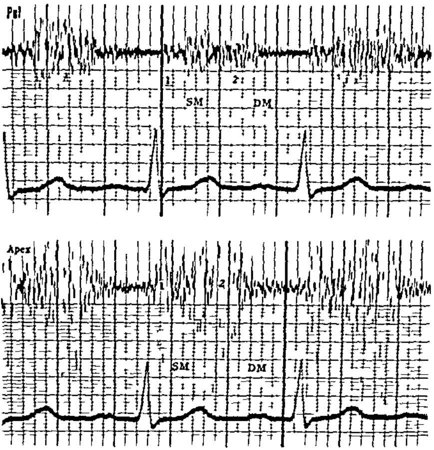


Figure 204 Continuous machinery type of murmur at the pulmonary area and a loud systolic and low frequency diastolic murmur at the apex

ventricular and left atrial enlargement (2) prominent pulmonary artery and large ascending aorta and aortic arch and (3) pulmonary vascular engorgement[98] (Figs 205 206) At fluoroscopy if the ductus is of an appreciable size the beat is hyperactive the increased amplitude of pulsations involves to an equal degree the left ventricle pulmonary artery and aorta Expansile pulsations are frequently seen in the engorged pulmonary vessels

The electrocardiogram like the radiogram may be almost entirely within normal limits in patients with a small ductus (Fig 207) If the shunt is large the increased left ventricular flow work is reflected in left ventricular hypertrophy with an occasional instance of P mitrale (Fig 208)

Cardiac catheterization (Fig 209) is barely justifiable today in patients with typical patent ductus arteriosus Earlier observations revealed the following data (1) The oxygen content of the pulmonary artery is increased at least 1 volume per cent over that of the right

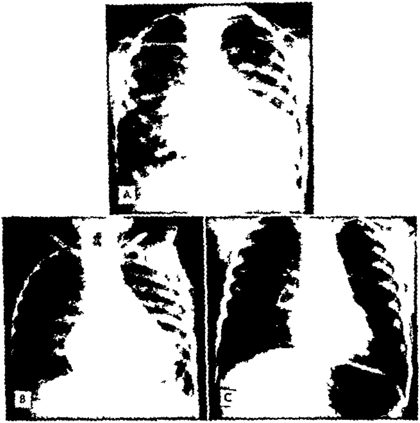


Figure 209 Three posteroanterior radiograms one belonging to a patient with a small patent ductus arteriosus (*C*) the second to a patient with medium-sized (*A*) and the third one with a large patent ductus arteriosus (*B*)

ventricle (2) Right ventricular and pulmonary arterial pressures are normal (3) The "pulmonary capillary" pressure is normal or slightly increased and the brachial arterial tracing shows a rapid upstroke time (4) Arterial saturation is normal (5) The pulmonary flow is usually at least twice the size of the systemic flow and the pulmonary resistance is normal or only slightly elevated (6) Occasionally the catheter passes from the pulmonary artery to the aorta (Fig 210) This occurs most commonly in the youngest patients with the largest ductus

In our hands angiocardiography has not been a particularly useful method of demonstrating a patent ductus arteriosus On the other hand retrograde aortograms may give a most graphic demonstration of the presence of a patent ductus Optimally the duct itself may be visualized (Fig 123 p 125) often the presence of a ductus can be assumed from the simultaneous opacification of the arch of the aorta and the pulmonary arterial tree

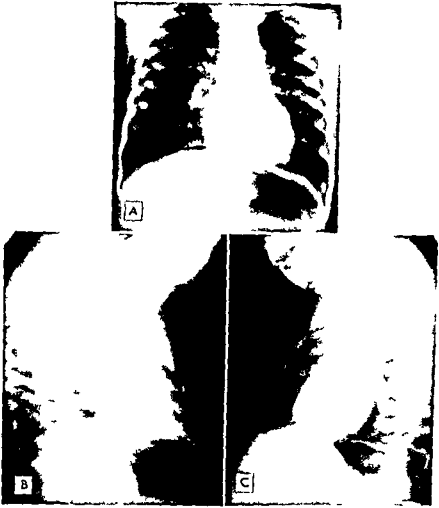


Figure 206 Posteroanterior and oblique radiograms in patient with small patent ductus arteriosus

Course and Prognosis

As mentioned earlier only rarely is a typical patent ductus arteriosus diagnosed within the first few months of life and practically never is the murmur recognized at birth The rare presence of a continuous murmur at birth usually means another probably more complex anomaly

Why a ductus arteriosus that presumably was present in intrauterine life and will again show evidences of patency at one or two years of age should not be detectable by auscultation at birth presents an interesting problem The answer may lie in the absence of a significant pressure gradient between the systemic and pulmonary circuits in other words the ductus may be anatomically open but there is no flow through

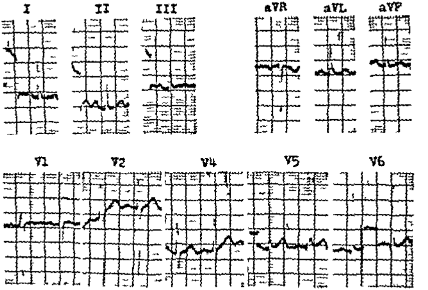


Figure 207 Electrocardiogram in a patient with small patent ductus arteriosus Note that the only abnormality is the presence of slightly tall voltages in the left chest leads

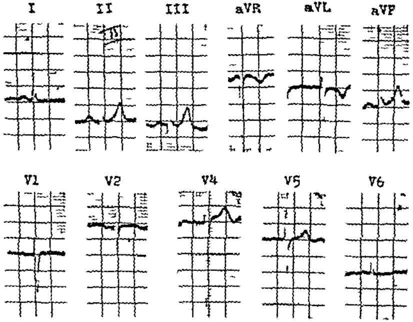


Figure 208 Electrocardiogram of a patient with large patent ductus arteriosus Note the presence of P mitrale in lead I and T wave changes in lead V_6

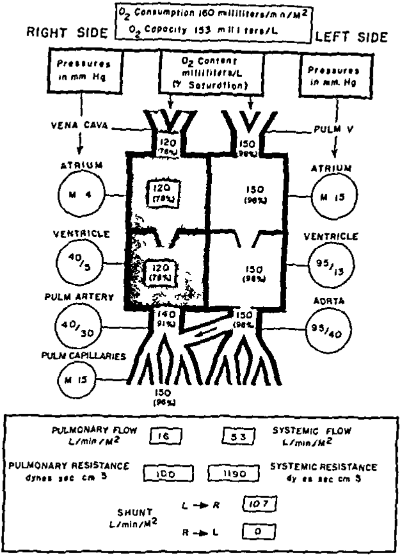


Figure 209 Diagrammatic presentation of cardiac catheterization data in patient with typical patent ductus arteriosus

it because the fetal type of pulmonary arterioles may represent a resist ance only slightly if at all lower than the systemic resistance In other cases the assumption has been made that a membrane obliterating the ductus has suddenly ruptured[331] In addition to these anatomic and physiologic explanations human factors consisting in the sometimes exceedingly difficult auscultatory conditions in the newborn may play some role in the late discovery of the typical murmur

Even if a murmur is discovered within the first few weeks or months of life it is seldom labeled as a continuous one Most often only a sys tolic component is recognized probably because only a systolic gradient



Figure 210 Passage of the catheter from pulmonary artery to aorta through a patent ductus arteriosus

between the two circuits exists By the end of the first or second year of life however in most cases of typical patent ductus arteriosus the characteristic murmur is noticeable The average patient with this anomaly is nearly asymptomatic throughout childhood As time goes on fatigue and breathlessness become increasingly noticeable and congestive failure may occur when he reaches the twenties or thirties The occurrence of pulmonary arterial hypertension with pulmonary vascular obstruction is fairly common in these older patients Shapiro and Keyes[309] found that a person with a patent ductus arteriosus who reaches his seventeenth birthday has only half the average life expectancy if the defect is not corrected by operation Individual exceptions to this rule are numerous and every cardiologist has seen patients with patent ductus arteriosus who in their fifties and sixties are completely asymptomatic

The great dangers for these patients are congestive failure and bacterial endocarditis It is possible that with the decline in incidence of the latter complication the figures of Shapiro and Keyes may no longer be valid It is my impression that rheumatic fever and rheumatic heart disease are also more common among patients with patent ductus arteriosus than among the average population

Differential Diagnosis

The diagnosis of typical patent ductus arteriosus is simple Gibson[130] stated that the clinical diagnosis of patent ductus arteriosus based prin

cipally on the presence of the characteristic murmur was correct in practically every instance of several thousand cases collected from all over the United States Other conditions with systolic and diastolic murmurs to be considered are as follows

Aortopulmonary Septal Defect An aortopulmonary septal defect is practically indistinguishable from a patent ductus arteriosus from the clinical viewpoint Cardiac catheterization aortography or both may furnish the necessary clue but exploratory thoracotomy should always be performed in any case

Coronary Arteriovenous Aneurysm A coronary arteriovenous aneurysm may be suspected if high frequency continuous murmurs are heard somewhere within the cardiac border but not at the second left interspace Cardiac catheterization may have to be performed to establish the diagnosis

Murmurs of Collaterals Murmurs of collateral vessels may be heard in patients with coarctation of the aorta or tetralogy of Fallot and may easily be identified by their location and character and the presence of the underlying malformation These murmurs are heard well throughout the back and usually at the second right interspace rather than being localized to the area of the ductus Also they lack the consistency of the patent ductus arteriosus murmur and vary considerably with respirations

Venous Hum A venous hum is perhaps most frequently confused with a patent ductus arteriosus Its evanescence the accentuation of the diastolic component its disappearance on compression of the ipsilateral jugular vein its accentuation if the neck is extended and the patient sits up and its virtual disappearance when he lies down usually allow an easy differential diagnosis

✓ **Ventricular Septal Defect and Aortic Regurgitation** The murmur of ventricular septal defect with aortic regurgitation is more to-and fro in character and its maximal intensity is at the lower left sternal border rather than at the second left interspace

Rupture of the Sinus of Valsalva If an aneurysm of the sinus of Valsalva ruptures into the right ventricle (ordinarily some time after puberty) the patient usually experiences the sudden appearance of congestive failure Physical examination reveals for the first time signs suggestive of a patent ductus arteriosus in a person without known heart disease This contrasts markedly with the slowly progressive course of patients with patent ductus arteriosus

Truncus Arteriosus The presence of arterial unsaturation the absence of a well defined main pulmonary artery segment and the common occurrence of right ventricular hypertrophy usually make the distinction between truncus arteriosus and patent ductus arteriosus fairly easy Also the continuous murmur is seldom localized to the second

left interspace but may be heard with equal intensity at the second right interspace and over the back

Treatment

The treatment of choice for typical patent ductus arteriosus is surgery An operation is recommended for every child with this condition who is seen at The Children s Medical Center in Boston This policy is based principally on two considerations one is the unlikelihood that any patient with a patent ductus arteriosus will be able to live a symptom free life for the average length of time and the other is the fact that normal circulation is completely restored by this procedure with a low mortality rate

It is easy to recommend surgery for patients who have symptoms However recommending an operation no matter how safe for a child who in the eyes of his parents is completely healthy is difficult The physician should be guided by his judgment of the individual situation He should try to assess the size of the ductus on the basis of heart size pulse pressure and presence or absence of an apical diastolic murmur Consideration should be given to the parents psychological make up Are they more able to withstand the emotional strain of an operation better than living with the threat of bacterial endocarditis for many years? A final most important consideration is the amount of surgical experience with this procedure in the community I believe that if one rationally examines all these factors and has a capable surgical team at ones disposal operation will be recommended for the vast majority of asymptomatic children as well as for those who have symptoms

The question—fortunately seldom confronting the pediatrician—of what to advise an adult with an asymptomatic ductus is an even more difficult problem I believe that not every one of these adults should be operated on the smallest ductus may perhaps be left undisturbed as long as the patient is asymptomatic This difference in attitude toward adults is explained by the slightly higher surgical risk that these patients present the sociologic implications of losing the head of the family and finally by the fact that the benignancy of the lesion has already been demonstrated to a certain extent by freedom from symptoms for several decades

Great technical strides have been made in the performance of this operation since the first successful ligation of a patent ductus arteriosus in 1939 by Gross [15] At present the members of our surgical service at The Children s Medical Center believe that division of the ductus is more satisfactory than ligation and is equally safe The principal advantage of dividing the ductus is the complete assurance against recanalization and subsequent development of bacterial endocarditis at least eight cases of recanalization of ligated ductus have been reported by Gibson [130] The mortality rate of division of patent ductus arteriosus is quoted by



Fig 211 A

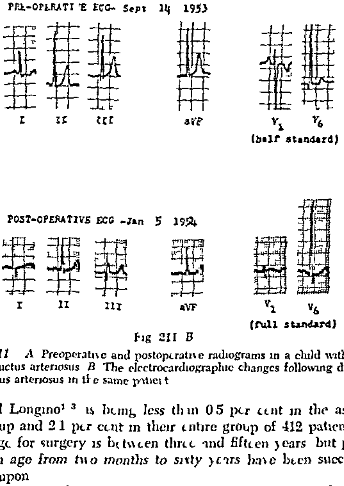


Fig 211 B

Figure 211 A Preoperative and postoperative radiograms in a child with division of patent ductus arteriosus B The electrocardiographic changes following division of patent ductus arteriosus in the same patient

Gross and Longino[1,3] is being less than 0.5 per cent in the asymptomatic group and 2.1 per cent in their entire group of 412 patients The optimal age for surgery is between three and fifteen years but patients *ranging in age from two months to sixty years* have been successfully operated upon

If bacterial endocarditis occurs it is preferable to eradicate the

infection medically first and to operate about six months after complete bacteriologic cure If the infection cannot be controlled by the most skillful use of antibiotics surgery may bring about the desired cure of the endocarditis

If the patient shows evidences of congestive failure obviously this should be brought under control before operation

The technical details of the operation and the preoperative and postoperative care are adequately covered in the appropriate surgical texts[151] On the whole the course of these patients after operation is remarkably smooth and in marked contrast to that following the repair of atrial septal defects is usually not plagued by either congestive failure or arrythmias The average stay in the hospital is two weeks or less

Postoperatively the heart diminishes in size pulse pressure becomes normal electrocardiographic and radiologic evidences of left ventricular hypertrophy disappear and pulmonary vasculature becomes normal (Fig 211) In the majority of instances no murmur is heard by the time the patient is discharged from the hospital Occasionally a child has a residual systolic murmur (grade 2 to 3) at the apex or the lower left sternal border most of these disappear within two years A stenotic (aortic or pulmonary) type of murmur may appear or as pointed out previously may in fact, have been present even before division of the ductus

SUMMARY

Typical patent ductus arteriosus can be diagnosed in the overwhelming majority of instances by the presence of a continuous murmur at the second left interspace Ancillary findings include radiologic and electrocardiographic evidences of a left to right shunt with increased left ventricular flow work Most patients are asymptomatic throughout childhood but with increasing years they may become incapacitated and may die in congestive failure or with bacterial endocarditis Surgical division a safe and successful procedure is recommended for all children with this anomaly

Patent Ductus Arteriosus with Pulmonary Hypertension (Atypical Patent Ductus Arteriosus)

CLINICAL PICTURE

The majority of these patients have moderate or severe symptoms Most of them are poorly developed and undernourished and are subject to frequent attacks of severe respiratory infection Spells of cyanosis are commonly noted and respirations are often rapid and labored A murmur has been noted in all of them but this is by no means the principal reason for their being brought to a cardiologist most of them are sick children indeed

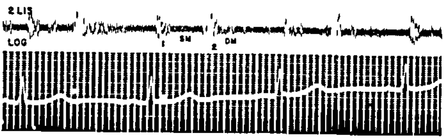


Figure 212 Phonocardiogram of patient with atypical patent ductus arteriosus Note the to-and fro rather than continuous character of the murmur Also the crescendic character of the systolic component is missing

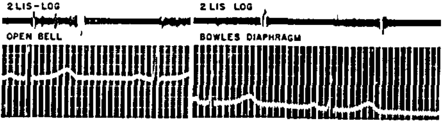


Figure 213 Phonocardiogram of patient with atypical patent ductus arteriosus Note the faint short early diastolic murmur

As a rule at physical examination these patients are thin and scrawny seldom do they rank above the tenth percentile on our standard developmental charts Redness of the fingers and toes is common Frank cyanosis and clubbing is present in less than 10 per cent of the cases if such is noted it may be either generalized or localized to the left hand and the lower half of the body The neck veins may be distended left chest deformity of moderate or severe degree is the rule The cardiac impulse is rocking and palpable at the xiphoid process as well as at the apex A thrill may be present at the upper left sternal border The systolic pressure is usually elevated with a widening of the pulse pressure In the occasional patient with congestive failure pulmonary rales are heard and the liver is enlarged but not pulsatile

At auscultation the dominant feature is the booming, narrowly split pulmonary closure The first and third heart sounds are not remarkable A grade 3 to 4 systolic murmur mostly crescendic in configuration is heard best at the second to third left interspace and transmits well throughout the precordium but not too clearly to the neck and the back. An early diastolic murmur is heard in about two thirds of the cases Commonly the systolic and diastolic elements do not form a continuous murmur a hiatus exists between the systolic murmur and the second sound giving it a to and fro character rather than a continuous one (Fig 212) Even if the systolic murmur does continue through the second sound the diastolic murmur may be short and thus again the criteria for a truly "continuous" murmur are not *completely fulfilled* (Fig 213) In a minority of cases only a systolic murmur at the second

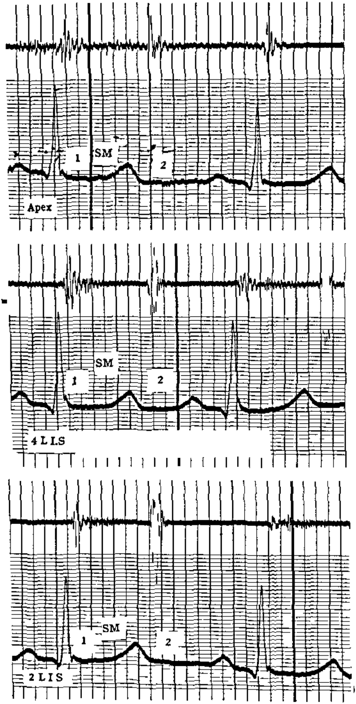


Figure 214 Phonocardiogram of patient with atypical patent ductus arteriosus Note the faint systolic murmur No definite diastolic murmur is noted

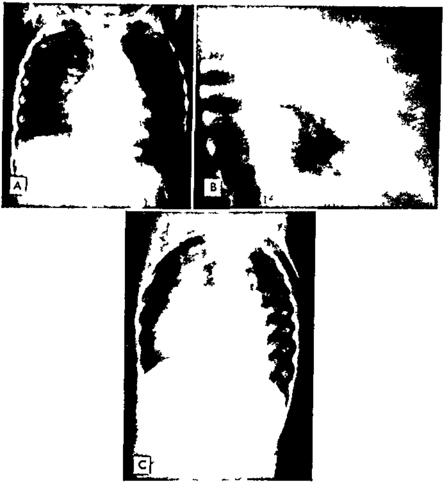


Figure 215 Posteroanterior and oblique radiograms of patient with atypical patent ductus arteriosus Note the prominent main pulmonary artery the absence of pulmonary plethora and the evidence of right ventricular enlargement in the left anterior oblique view

left interspace is heard (Fig 214) In addition to the murmur at the second left interspace apical systolic and mid diastolic murmurs are also common

Roentgenologic examination always reveals a grossly abnormal cardiac silhouette In addition to the enlargement of the left ventricle left atrium and pulmonary artery there is definite and even dominant right ventricular enlargement as well (Fig 215) Frequently in addition to the active pulmonary vascular engorgement inherent in a left to-right shunt of some magnitude there may be evidences of passive congestion In the most advanced cases with pulmonary vascular obstruction the marked contrast between engorged hilar vessels and ischemic peripheral lung fields can be seen

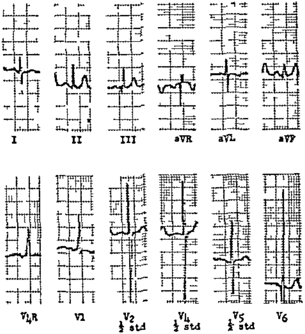


Fig 216 A

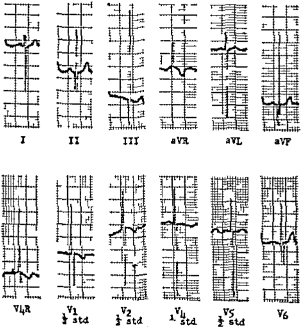


Fig 216 B

Figure 216 Electrocardiograms of patients with atypical patent ductus arteriosus *A* Mild right ventricular hypertrophy with definite evidence of left ventricular hypertrophy *B* Exclusively right ventricular hypertrophy

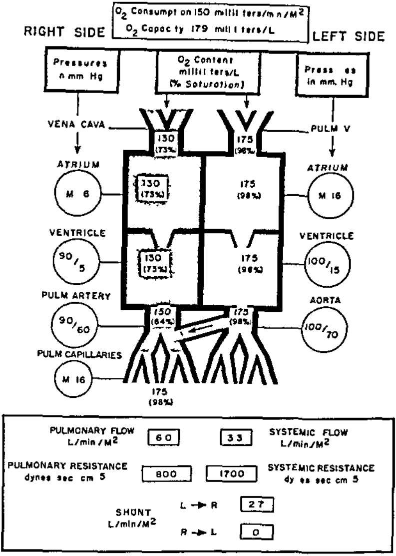


Figure 217 Diagrammatic representation of cardiac catheterization data in patient with atypical patent ductus arteriosus

The electrocardiogram frequently reflects the increased pressure work of the right ventricle by showing the presence of combined or even pure right ventricular hypertrophy with or without incomplete right bundle branch block (Fig 216)

Cardiac catheterization (Fig 217) reveals the following (1) the oxygen content of the pulmonary artery is increased at *least 1 volume* per cent over that of the right ventricle Occasionally probably because of pulmonary regurgitation the oxygen content of the right ventricular outflow tract may be significantly higher than that of the body of the

right ventricle or the right atrium (2) Right ventricular and pulmonary arterial hypertension of moderate to severe degree is present (3) The "pulmonary capillary" pressure is increased and there is a normal or regurgitant type of brachial arterial tracing (4) Moderate arterial oxygen unsaturation exists in 20 to 25 per cent of the patients This is particularly true in the samples obtained from the femoral artery in cases of a reversed ductus that is a ductus carrying blood from the pulmonary artery to the aorta If the femoral arterial blood has an oxygen content of at least 1 volume per cent less than that of the fully saturated right brachial artery the diagnosis is unquestionably reversed ductus (5) The pulmonary flow is moderately or sizably larger than the systemic flow the pulmonary resistance is always increased to a greater or lesser degree (6) The catheter frequently passes from the left pulmonary artery through a large ductus into the descending aorta

Analysis of the data obtained at cardiac catheterization reveals that in most infants and children with atypical ductus the pulmonary arterial hypertension is the result of a large increase in pulmonary blood flow with only a moderate increase in the pulmonary vascular resistance In a minority of the children and many adults however there may be pulmonary hypertension with only a small increase in pulmonary flow and a marked elevation of pulmonary vascular resistance This latter group can be recognized clinically by the presence of cyanosis electrocardiographic evidences of pure right ventricular hypertrophy and at catheterization an identical mean pressure in the pulmonary and systemic arteries This combination of symptoms is much more likely to occur in late adult life than during childhood

Angiocardiography (with injection of the dye into the right side of the heart) may be extremely helpful in revealing the ductus and the descending aorta in patients with significant arterial unsaturation On the other hand patients without arterial unsaturation can be studied more profitably by retrograde aortography

Course and Prognosis

Although in later years pulmonary arterial hypertension with pulmonary vascular obstruction may develop in patients with typical patent ductus arteriosus on the whole this is a rather unusual happening The patients being discussed here all had significant pulmonary hypertension in childhood, half of them in infancy Whereas the late development of pulmonary vascular disease in the typical case of patent ductus arteriosus may be attributable to the prolonged effects of a large pulmonary blood flow[92, 351] the presence of high pulmonary arterial pressure in infancy or early childhood must be explained on a different basis Civin and Edwards[81] stated that in these cases the size of the ductus for teleologic reasons necessitates a narrowing of the pulmonary vascular bed in order to maintain an adequate pressure in the systemic circuit as a matter of

fact they demonstrated the persistence of the fetal type of pulmonary arterioles with narrow lumens in some of their infants with the largest ductus According to this theory then in patients with large aortopulmonary communications the pulmonary arterioles do not undergo the usual postnatal change consisting in an increase in the lumen the increased pulmonary vascular resistance thus maintained helps to reduce the flow through the duct and to maintain an adequate systemic output Attractive as this hypothesis may be I do not believe that it explains the pulmonary hypertension in all the patients in the group Actually in some instances a ductus of only moderate size is accompanied by significant pulmonary vascular obstruction whereas in others even though the ductus is very large the pulmonary vasculature is entirely normal for the patient s age both anatomically and physiologically

Thus it seems that patients having an atypical ductus or a ductus with pulmonary hypertension do not represent a physiologically homogeneous group Rather there are two groups The first group is made up of infants with large patent ductus tremendously increased pulmonary blood flow and only slightly increased resistance there is some evidence that after a few years with only a moderate increase in the actual pulmonary arterial pressure but with marked progression of the pulmonary vascular resistance the left to-right shunt may diminish and pulmonary vascular obstruction may appear in some of these infants These patients like those with large ventricular septal defects have a stormy infancy replete with episodes of congestive failure pulmonary edema and attacks of pneumonia This difficult period is followed by a relatively symptom free childhood finally pulmonary vascular obstruction occurs with cyanosis in adolescence and death in early adult hood At one time or another these patients may present auscultatory phenomena typical of patent ductus arteriosus electrocardiographic evidences of left ventricular hypertrophy may exist transiently only to give way to evidences of combined ventricular hypertrophy At cardiac catheterization the pressure in the pulmonary artery is significantly elevated but not to the height of the systemic pressure pulmonary resistance is usually less than systemic resistance

The second group is made up of patients who have a large or moderate sized ductus with marked pulmonary vascular obstruction that may well be congenital These people do not go through the same period of increased pulmonary blood flow as those in the first group *they are cyanotic from birth and evidences of pulmonary vascular* obstruction dominate their entire clinical picture Strangely their life expectancy may not be any shorter than that of the first group but their symptoms are more uniform throughout life These are the patients in whom the auscultatory findings are the least typical in whom electrocardiographic evidences of pure right ventricular hypertrophy are encountered and in whom, at cardiac catheterization the highest pulmonary

arterial pressures associated with calculated pulmonary resistances equal to or higher than systemic resistance are encountered

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of atypical patent ductus arteriosus is one of the most difficult and most crucial encountered in pediatric cardiology *In my opinion an atypical patent ductus arteriosus should be excluded by cardiac catheterization in every case of a left to right shunt with pulmonary arterial hypertension if evidences of left ventricular hypertrophy are present and particularly if the pulse pressure is wide* If catheterization studies fail to furnish an unequivocal answer a retrograde aortogram or even surgical exploration should be contemplated

If patent ductus arteriosus with pulmonary arterial hypertension is diagnosed on clinical grounds often the abnormal auscultatory roentgenographic or electrocardiographic features suggest the presence of an additional congenital malformation Although accompanying congenital defects notably atrial or ventricular septal defect pulmonary stenosis aortic stenosis and so forth have been described by several authors [153, 174] the presence of such abnormalities should be diagnosed only after thorough studies In my experience most atypical" features can be explained solely on the basis of the ductus with pulmonary arterial hypertension Furthermore for practical purposes the presence of an additional congenital malformation should not make any difference from the therapeutic viewpoint except in the extremely rare instance in which a tetralogy of Fallot is associated with a patent ductus arteriosus The association of pure pulmonary stenosis or aortic stenosis with patent ductus arteriosus does not represent a contraindication to operation neither do septal defects

The principal lesions to be considered in the differential diagnosis are as follows

Total Anomalous Pulmonary Venous Drainage The marked cardiac enlargement with congestive failure the severe and pure right ventricular hypertrophy without much evidence of pulmonary arterial hypertension and the characteristic roentgenogram usually enable the cardiologist to identify total anomalous pulmonary venous drainage Occasionally however only the demonstration of systemic arterial blood in the right atrium at catheterization furnishes an unequivocal answer to the problem

Atrial Septal Defect or Partial Pulmonary Venous Anomaly Atypical patent ductus arteriosus resembles atrial septal defect or partial pulmonary venous anomaly only from the radiologic viewpoint. The similarity of the two in terms of prominent main pulmonary artery with right ventricular hypertrophy and pulmonary vascular engorgement may be striking Consideration of other factors however particularly the presence of a widely split, but not accentuated pulmonary second sound,

the selective right ventricular enlargement shown in the roentgenogram and the marked incomplete right bundle branch block without evidences of left ventricular hypertrophy as indicated by the electrocardiogram usually enables the experienced observer to identify a communication at the atrial level

Large Ventricular Septal Defect Large ventricular septal defect (with or without pulmonary stenosis or aortic regurgitation) may be impossible to separate from atypical patent ductus arteriosus without catheterization or even thoracotomy Localization of the murmur at the lower left sternal border the absence of the early diastolic components and the presence of normal pulse pressure and of pure right ventricular hypertrophy all tend to favor a ventricular defect However even the most experienced observers are likely to make serious mistakes Hence it cannot be emphasized too often that if this particular point in differential diagnosis is seriously considered catheterization should be carried out at the appropriate time particularly if the patient is not doing well

Truncus Arteriosus In addition to the points enumerated on page 352 it should be mentioned that atypical ductus arteriosus if accompanied by severe enough pulmonary arterial hypertension to give rise to cyanosis usually is not accompanied by a continuous murmur as loud and as ubiquitous as is seen with truncus arteriosus

Aortopulmonary Fenestration Aortopulmonary fenestration was discussed on page 352

Rupture of the Sinus of Valsalva See page 352.

TREATMENT

The treatment of patients with patent ductus arteriosus and pulmonary arterial hypertension cannot be summarized as easily as that of patients with typical patent ductus

Patients who have large left to right shunts and only a slight increase in pulmonary resistance unquestionably should be subjected to operation these patients may benefit tremendously from surgery It should be remembered however that the mortality rate for these patients some of them small babies is considerably higher than for those with asymptomatic ductus No large enough series is available to provide statistically meaningful figures however usually a mortality rate of 5 to 10 per cent should be quoted Furthermore in view of the potential progression of the pulmonary vascular changes operation is indicated as soon as the diagnosis is made Usually this does not seem like a difficult decision since as mentioned earlier, these children are sick and need help

The decision to operate on a patient belonging to the group in whom the pulmonary arterial hypertension is extremely severe and is principally due to marked pulmonary vascular obstruction without a significant left to right shunt is much more difficult to make Without

operation these patients cannot look forward to a normal life Death in five ten or fifteen years seems a foregone conclusion At the same time operation carries at least a 50 per cent risk Furthermore even if patients do survive the operation the chances of reducing the established pulmonary vascular obstruction are remote

The literature is filled with glowing reports of a postoperative decrease in pulmonary arterial pressure in patients operated on for patent ductus arteriosus but careful study usually indicates clearly that the pulmonary arterial pressure drop paralleled the drop in pulmonary blood flow I am not aware of any well substantiated case report in which an unequivocal and significant decrease in pulmonary arterial resistance was obtained by dividing the patent ductus arteriosus So far even though some of these patients with severe even maximal pulmonary vascular obstruction have survived the operation improvement has not followed very clearly It is hoped that the vascular obstruction will not progress any further but even this is more a matter of wishful thinking than proved fact

The medical support needed by these patients before during and after operation is appreciable Since congestive failure and pulmonary infections are common the need for preoperative anticongestive measures and antibiotics is great The use of spasmolytics such as Priscoline to reduce pulmonary obstruction has been tried In our hands the results on the whole have been discouraging

SUMMARY

Atypical patent ductus arteriosus should be suspected in every patient who has evidences of a left to right shunt with pulmonary arterial hypertension These patients are commonly subject to episodes of congestive failure and attacks of pneumonia physically they are greatly underdeveloped At auscultation in the great majority of instances a systolic murmur is present at the upper left sternal border early diastolic murmurs are also present in more than half of the patients The pulse pressure may be widened Electrocardiograms and roentgenograms show combined ventricular hypertrophy Cardiac catheterization is indicated for every patient in whom such a lesion is seriously suspected

AORTOPULMONARY FENESTRATION

It may be worth while to mention briefly a rare congenital anomaly which closely resembles patent ductus arteriosus in all respects except operability namely aortopulmonary fenestration or aortic septal defect

Embryologically this is the result of the incomplete development of the spiral ridges dividing the bulbus cordis clearly this should result in a persistence of the fetal communication between the two halves of the bulbus

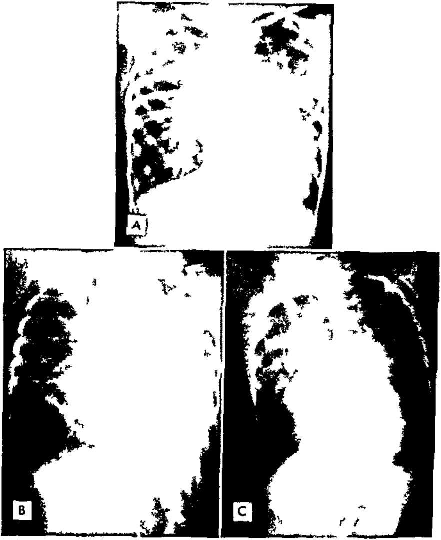


Figure 218 Posteroanterior and oblique radiograms of patient with aortopulmonary fenestration Note the similarity between these films and those of patient with large patent ductus arteriosus

Anatomically the defect varying in diameter from 5 to 30 mm [150] is located just above the aortic valve Sometimes it is practically inseparable from the coronary arteries in other instances it is a few millimeters above them

The physiologic effects of an aortopulmonary fenestration are identical with those of a patent ductus arteriosus because of the gradient between systemic and pulmonary resistances a shunt from the aortic to the pulmonary side occurs The size of the defect and the ratio between the two resistances are the principal factors determining the magnitude of the left to right shunt

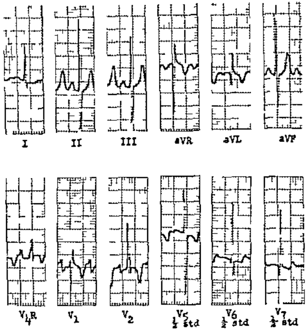


Figure 219 Electrocardiogram of patient with aortopulmonary fenestration Note the presence of combined ventricular hypertrophy

Clinical Picture

For all practical purposes the clinical picture cannot be distinguished from that of a large patent ductus The only clue to the fact that this is not a clear cut ductus arteriosus is that the murmur is usually somewhat atypical and is situated lower and more medially than is customary

The roentgenologic picture (Fig 218) and electrocardiographic findings (Fig 219) are characteristic of a large patent ductus arteriosus perhaps associated with pulmonary hypertension even cardiac catheterization studies may not differentiate between the two entities (Fig 220) Gasul and his associates[1 7] reported one case in which differentiation was possible by means of retrograde aortography

Course and Prognosis

The natural history of the anomaly cannot be outlined with any certainty because of the paucity of reported cases Survival varies from three days reported by Hektoen[163] to forty eight years reported by Moorhead and Smith[45] Death usually occurs from congestive failure probably associated with pulmonary vascular obstruction although these patients should be equally susceptible to bacterial endocarditis as are patients with patent ductus arteriosus

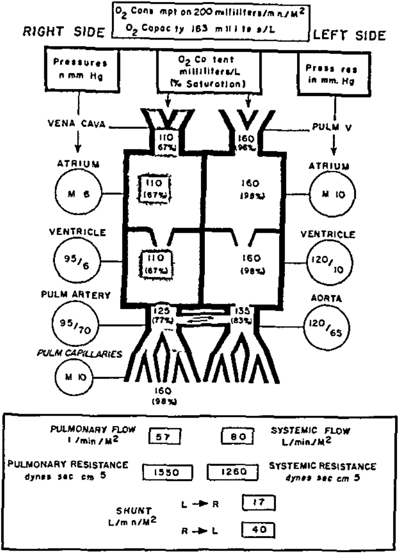


Figure 220 Diagrammatic presentation of cardiac catheterization data of a patient with aortopulmonary fenestration.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of aortopulmonary fenestration from large patent ductus arteriosus is practically impossible by clinical means and extremely difficult by cardiac catheterization or angiography it can be accomplished with certainty only at thoracotomy

TREATMENT

Surgical treatment of the aortopulmonary septal defect is exceedingly difficult Only two or three successful operations have been reported

so far in the literature. However, accurate diagnosis of the condition is possible only at the operating table. The usual approach toward division or ligation of a patent ductus arteriosus is pursued. When the ductus is exposed, only a ligamentum arteriosum is found, the manual constriction of which does not appreciably change the dynamics of the heart or eliminate the loud thrill in the pulmonary artery. Under these circumstances the surgeon's exploring finger will discover that the maximal intensity of the thrill is further toward the heart, just above the semilunar valves. Then, if the pericardium is opened, the aortic septal defect can almost always be located at its usual place. The difficulty in interrupting this structure arises from the short distance between the two great vessels and the proximity of the defect to the coronary arteries. Nevertheless, with the appropriate technique, occasionally a successful operation can be performed.[1, 9]

Summary

Aortopulmonary fenestration is a communication between the two great arteries just above the semilunar valves. The diagnosis should be suspected in patients who have symptoms of large and somewhat atypical ductus, but it can be proved only at operation. Surgical treatment is extremely difficult because of the anatomic situation, but a few successful operations have been reported.

Ruptured Aneurysm of the Sinus of Valsalva

Aneurysmal dilatation of the aortic sinus is the result of a congenitally imperfect fusion of the bulbar ridges and the ventricular septum. It is a developmental disturbance only slightly less profound than that leading to a defect of a membranous ventricular septum; as a matter of fact, in my experience and according to reports from the literature, the two lesions frequently occur together.[61, 333] The aneurysmal dilatation apparently never involves the left cusp and is most frequent in the right cusp. Whether the communication frequently existing between this sinus and the right side of the heart is congenital or acquired is a matter of considerable argument; according to the excellent review of Jones and Langley,[179] the communication is acquired in about three fourths of the instances. The opening of the aortic sinus usually leads into the right ventricle from the right cusp and only occasionally into the right atrium from the posterior cusp.

Clinical Picture

A congenital communication between the sinus of Valsalva and the right ventricle cannot be distinguished with certainty clinically or at cardiac catheterization from a large ductus or a ventricular septal defect with aortic regurgitation. Although retrograde aortography may furnish the necessary clues for accurate recognition, only at thoracotomy can the

definite diagnosis be made In cases in which the aneurysm ruptures later in life the diagnosis may be suspected if evidences of congestive failure associated with features characteristic of a large ductus develop suddenly within a matter of days or weeks An aneurysm of the sinus of Valsalva with no communication to the right side of the heart and without associated defects does not give rise to signs or symptoms

Course and Prognosis

Patients in whom this communication is congenital may lead relatively normal lives throughout childhood however death has been reported in early infancy Patients in whom the communication is acquired usually die within a few months after the onset of congestive failure

Treatment

No surgical treatment has been devised so far Medical regimen should be aimed at controlling evidences of heart failure and infection

Summary

Aneurysm of the aortic sinus is in itself an asymptomatic condition Only if a communication develops between the sinus of Valsalva and the right side of the heart do symptoms and signs strongly resembling those of patent ductus arteriosus become manifest Only if the communication is acquired can the diagnosis be suspected preoperatively principally on the basis of the course of the disease The prognosis is poor and no treatment is available at present

Persistent Truncus Arteriosus

Incidence

Truncus arteriosus is not a common lesion At The Childrens Medical Center we have had eleven children with this condition among our 577 proved cases of congenital heart disease Abbott reported twenty one in her series of 1000 cases of congenital malformations of the heart

Anatomy

Complete failure of development of the bulbar ridges results in the persistence of the fetal common arterial trunk Since these ridges are also necessary to complete the development of the ventricular septum persistent truncus arteriosus is always associated with a defect of the membranous septum and invariably communicates with both ventricles Frequently the entire ventricular septum is absent The number of semilunar valves varies from two to four The ductus is absent by definition The entire circulation including the systemic arteries the pulmonary arteries and the coronary arteries is supplied from this common arterial trunk

Whereas in the majority of instances the aorta with its tributaries and the coronary arteries are functionally adequate the state of the pulmonary vessels may vary considerably In some cases normal sized pulmonary arteries originate directly from both sides of the common trunk in other cases one or even both pulmonary arteries are absent and the pulmonary circulation is taken over entirely by the bronchial arteries I have even seen an instance in which the right pulmonary artery was given off by the common arterial trunk while the left pulmonary artery originated from the right ventricle

PHYSIOLOGY

Since blood leaving the heart through one common exit represents a practically complete mixture of the pulmonary (oxygenated) and systemic (unoxygenated) return the arterial saturation of the truncus blood depends on the ratio of the systemic to the pulmonary flow If the pulmonary arteries are adequate in size and the pulmonary circulation therefore large the truncus blood will be relatively highly saturated Conversely if the pulmonary circulation is inadequate either because of small pulmonary arteries or pulmonary vascular obstruction the oxygen content of the blood in the common trunk will be nearly the same as that in the vena cava The systolic pressure in the two ventricles is equal and identical with that obtained from the aorta The pulmonary arterial pressure is variable The oxygen content in the two ventricles the aorta and the pulmonary arteries is identical

CLINICAL PICTURE

The clinical picture varies tremendously depending on the status of the pulmonary arteries A patient with normal sized pulmonary arteries and normal pulmonary resistance is practically acyanotic the clinical picture resembles that of a large ventricular defect or single ventricle On the other hand a child with hypoplastic pulmonary arteries or pulmonary vascular obstruction may exhibit the worst degree of cyanotic congenital heart disease

Most of the patients are known to have had heart disease since birth Their principal symptoms are dyspnea and fatigue with or without evidences of congestive failure At physical examination these children are poorly developed most of them rank well below the third percentile on our development charts and none of them ranks above the twenty fifth percentile line Prominence of the left side of the chest is common a thrill is usually palpable at the second left sometimes at the second right interspace Cyanosis and clubbing vary considerably from patient to patient One infant has been observed for almost two months in the wards at The Children s Hospital without definite evidence of cyanosis The blood pressure is usually normal although a person who has large

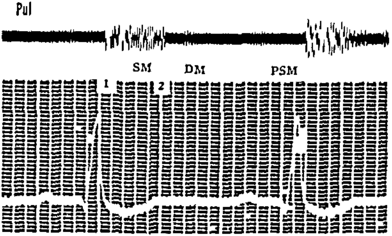


Figure 221 Systolic and diastolic murmurs in the pulmonary area in a patient with truncus arteriosus Note the decrescendo shape of the systolic murmur the faintness of the diastolic murmur and the presence of the presystolic murmur

pulmonary arteries may have a wide pulse pressure Hepatomegaly and rales are noted in patients who have congestive heart failure

The first heart sound is normal or accentuated Usually the second sound is accentuated or even booming The expected narrowness of the second sound is not observed with any regularity More than half of our patients had a continuous murmur at the second left or second right interspace This murmur is similar to the machinery murmur heard in patent ductus arteriosus except that its point of maximal intensity is not always at the pulmonary area the systolic component is not always crescendic and the intensity of the murmur varies with respirations (*Fig* 221) *Loud grade 4 systolic murmurs were heard at the lower left* sternal border or the second right interspace in all the patients who did not have a continuous murmur Apical mid diastolic and presystolic murmurs were also present in a minority of the patients

The electrocardiogram is variable Most patients show combined ventricular hypertrophy but occasionally pure left or right ventricular hypertrophy is present P pulmonale is never marked

The roentgenologic picture (Fig 222) is characterized by the ab sence of a normal main pulmonary artery segment at the usual place. The supracardiac shadow consists of one large vessel usually but not invariably to the left of the esophagus Cardiac enlargement is moderate or severe and usually involves both ventricles Pulmonary ischemia or unilateral or even bilateral pulmonary plethora may be noted depend ing on the size of the pulmonary arteries (Fig 223)

Cardiac catheterization (Fig 224) reveals the following (1) There is an increase in oxygen content at the right ventricular level resulting in a uniform oxygen content in the right ventricle systemic artery and pulmonary artery (2) The systolic pressure is the same in the right



Figure 222 Posteroanterior and left anterior oblique radiograms of patient with common truncus arteriosus Note the absence of a main pulmonary artery the large aorta left and right ventricular enlargement in the left anterior oblique view and the pulmonary plethora in the posteroanterior view

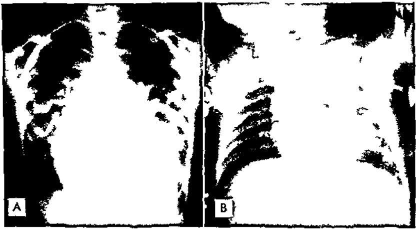


Figure 223 Posteroanterior radiograms of two patients with truncus arteriosus *A* Marked pulmonary plethora in one patient *B* Pulmonary ischemia in the second patient

ventricle as in the systemic artery with identical or reduced pulmonary arterial pressure (3) The pulmonary capillary" pressure is normal or slightly increased and the brachial arterial pressure curve is relatively normal (4) There is arterial unsaturation of variable degree (5) A left to right or right to left shunt is present at the ventricular level depending on the relationship of the resistances in the two circuits (6) The catheter cannot enter the pulmonary artery directly from the ventricle

Angiocardiographic studies are particularly apt to demonstrate graphically the anatomic situation (Fig 225) In many respects this tool is superior to the catheter in the diagnosis of common truncus arteriosus Optimally both examinations should be performed

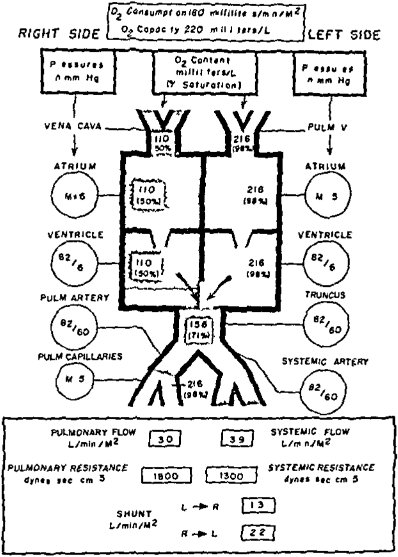


Figure 224 Diagrammatic presentation of cardiac catheterization findings in patient with common truncus arteriosus

Course and Prognosis

The natural history of the disease is variable Most of the patients die in infancy four of our eleven died between the ages of three and six months Our oldest living patient at this time is seventeen years old According to the literature many patients live to the age of twenty or thirty The immediate cause of death is usually congestive failure although pneumonia has contributed significantly to the death of our small infants One of our patients was severely ill with bacterial endocarditis another was successfully treated for a brain abscess



Figure .25 Angiocardiogram in left anterior oblique view in a patient with common truncus arteriosus Note the wide single supracardiac shadow

Differential Diagnosis

Persistent truncus arteriosus may have to be distinguished from other types of cyanotic or acyanotic congenital heart disease depending on the status of the pulmonary circulation

In the absence of cyanosis large ventricular septal defect and patent ductus arteriosus should be considered in the differential diagnosis A large ventricular septal defect is similar to truncus arteriosus in that both entail a communication at the ventricular level and both are commonly associated with pulmonary arterial hypertension The differences are discussed on page 320 The differential diagnosis of patent ductus arteriosus and persistent truncus arteriosus is discussed on page 352

In patients who have truncus arteriosus with cyanosis tetralogy of Fallot and tricuspid atresia should be considered in the differential diagnosis

In a patient with extreme tetralogy of Fallot with pulmonary atresia the pulmonary circulation flows through bronchial arteries or a patent ductus arteriosus from the aorta rather than through a small pulmonary artery from the common arterial trunk Often this differentiation is not possible except at postmortem examination at other times the typical bronchial arterial type of pulmonary vasculature identifies the condition radiologically Furthermore patients with extreme tetralogy of Fallot often have no murmur whatsoever whereas all the patients with a

Table 29

DIFFERENTIAL DIAGNOSIS OF COMMUNICATIONS BETWEEN GREAT VESSELS

	CLINICAL PICTURE													PHYSIOLOGY					COURSE & REMARKS
	M				X-RAY					E				SHUNT		PRESSURE			
PD	[illegible]	[illegible]	[illegible]			VE	[illegible]	PA		[illegible]	RV			→	→			[illegible]	
TYPICAL				APICAL HUMM						0			—					AR	[illegible] CHILDHOOD [illegible] DEATH [illegible]
TYPICAL	↔	f		EARL BLOW D	DR										—			DH AR	SICK INFANTS CHILDREN [illegible] MARKED [illegible] DE TH [illegible]
AORTO PULMONARY FENESTRATION	↔			EARL BLOW f						0									SAME ABOVE
RUPTURED [illegible] OF VALSALVA												+							RUPTURE [illegible] ACT LIKE LARGE [illegible] ACQUIRED, SUDDEN ONSET OF S, AFTER [illegible] EARL DEATH
TRUNCUS ARTERIOSUS						+													CLINICAL PICTURE DEPENDS ON PULM FLOW OR [illegible] MAY LIVE TO ADOLESCENCE

See Table 28 (p 303) for explanation of symbols

truncus arteriosus have a continuous or at least a systolic murmur Finally patients with extreme tetralogy of Fallot may be very cyanotic in spite of the presence of a continuous murmur originating from a relatively small bronchial artery In contrast, in patients with a true truncus arteriosus a continuous murmur implies an appreciable sized pulmonary artery and thus cyanosis though definite is of a lesser degree

Tricuspid atresia associated with an underdeveloped right ventricle may also be confused with true truncus arteriosus The question arises in a patient with cyanotic heart disease in whom the electrocardiogram reveals left ventricular hypertrophy Clinical differentiation may be possible on the basis of the severe and exclusive left ventricular hypertrophy with left axis deviation usually noted in tricuspid atresia In other instances only angiocardiography enables the cardiologist to make the diagnosis in vivo

Treatment

Only medical therapy (antibiotics and anticongestive measures) is available for the patients who have an adequate pulmonary circulation If on the other hand the pulmonary arteries are small or defective a shunt procedure may be successfully performed It must be remembered however that the operative mortality is high and the number of satisfactory results are few and far between Consequently surgery is to be recommended only for cyanotic patients in this group whose limitations are really maximal

SUMMARY

True truncus arteriosus is a large common vessel overriding both the left and the right ventricles. There may be severe or mild arterial unsaturation depending on the size of the pulmonary artery originating from this common trunk. The characteristic radiologic picture and the presence of a continuous murmur in a patient with cyanosis should suggest this entity. Angiocardiography with or without catheterization usually establishes the diagnosis. Surgical treatment may be available for patients with diminished pulmonary blood flow, but the mortality rate is high.

Valvular and Vascular Lesions with a Right-to-Left Shunt or no Shunt at All

COARCTATION OF THE AORTA

INCIDENCE

Coarctation of the aorta is a common congenital anomaly It is the third most common lesion in Abbott's report sixth most common in Wood's series and fifth in our own group of patients Males are more commonly affected than females On the whole this anomaly probably represents slightly more than 10 per cent of all congenital cardiac abnormalities

ANATOMY

Coarctation is a narrowing of the lumen of the aorta usually occurring beyond the origin of the left subclavian artery at the insertion of the ligamentum arteriosum Rare instances of coarctation higher in the aortic arch (even proximal to the innominate artery) [33] or far along the descending aorta have also been described

The etiology of coarctation of the aorta was thought to depend on the closure of the ductus arteriosus and the constriction resulting therefrom [331] Recent evidence points away from this explanation however Edwards stated that the lesion is due to an intrinsic defect in the structure of the medial layer of the aortic wall [107] It is my distinct impression that coarctation of the aorta is not of equal severity at birth in all patients and that in fact in many instances it may represent a gradually progressive lesion Whether the collaterals necessary for maintenance of an adequate postnatal circulation are developed during intrauterine life or not is a matter of lively controversy the available evidence favors the presence of these structures in the fetus The collateral channels bring blood to the lower half of the body by bypassing the aortic block In essence both subclavian arteries are the principal sources of the collaterals The right subclavian communicates to the intercostals via the vertebral arteries and the spinal artery on one hand and through the cer

vical and scapular branches on the other The left subclavian artery if proximal to the coarctation feeds blood to the lower half of the body through the internal mammaries and the intercostals

PHYSIOLOGY

The principal physiologic problem in coarctation of the aorta is the maintenance of adequate blood flow and pressure in the lower half of the body Apparently this is achieved by means of three adaptive mechanisms One is the elevation of the systolic pressure in the proximal aortic segment The second is vasoconstriction in the arterioles to maintain a high diastolic pressure The third is the bypassing of the aortic block by means of collaterals or a ductus arteriosus opening distal to the coarctation By utilizing one or more of these three mechanisms the mean pressure in the lower extremities is maintained at a level of about 60 mm of mercury the minimal pressure needed for adequate kidney function If the block is too complete or if the compensatory mechanisms are not sufficient, survival is not possible

A classification of coarctation of the aorta into "adult" and "infantile" types based on the location and length of the coarcted segment, is meaningless in view of present day physiologic and therapeutic concepts It is well known that the so called adult type (a localized constriction just opposite the insertion of the ductus arteriosus) may be associated with a patent ductus supplying the lower half of the body with blood and may cause death in congestive failure in infancy On the other hand the so called infantile type (a long narrow segment between the left subclavian and the ligamentum arteriosum) may be associated with relative longevity Also from the clinical viewpoint the adult type may give rise to severe though reversible congestive failure in infants whereas the infantile type can be completely asymptomatic for several decades

A physiologically as well as clinically more meaningful classification to be followed in this text divides coarctations of the aorta into those in which the left ventricle (directly or by means of collaterals) supplies the entire systemic circuit with blood (coarctation of the aorta with systemic left ventricle) and those in which the left ventricle supplies only the upper half of the body while the parts below the coarctation are supplied with blood from the right ventricle through a ductus arteriosus (coarctation of the aorta with systemic left and right ventricles)

COARCTATION OF THE AORTA WITH SYSTEMIC LEFT VENTRICLE

Simple Coarctation

CLINICAL PICTURE

Most children with coarctation of the aorta are asymptomatic the condition is usually first recognized by the school physician or the pedi

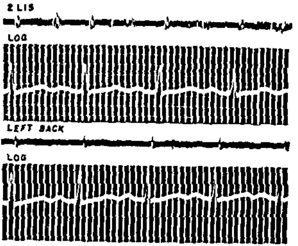


Figure 226 Phonocardiogram of patient with coarctation of the aorta Note the *stenotic* type of systolic murmur at the second left interspace and the back and the long diastolic murmur over the back

atrician when a routine examination reveals a murmur or the presence of arterial hypertension Once in a while close questioning of the parents elicits complaints consisting of pains in the calves on running occasional fatigue headaches and nosebleeds It is interesting that asymptomatic coarctation of the aorta is rarely discovered in infants it is usually diagnosed between the ages of four and five years or even later

At physical examination[4] these children (generally boys) are usually well developed and well nourished some of them are extremely athletic Occasionally one is struck by the broad shoulders and well developed upper extremities in contrast to the relatively narrow hips and thin legs Cyanosis and clubbing are never present, and there are no evidences of congestive failure Frequently a thrill is palpable at the suprasternal notch where the carotid pulsations are unusually powerful Mild left chest deformity may be present the impulse is heaving and is maximal at the cardiac apex In older children the dilated and tortuous intercostal arteries may be palpable over the back when the patient leans forward[7] The radial arteries are powerful especially on the right The femoral pulses are either absent or weaker and later than the radial pulses Blood pressure determinations (with proper sized cuffs) show the systolic pressure of the arms to be higher than that of the legs whereas the diastolic pressures are equal This relative hypertension of the upper extremities is the pathognomonic feature of coarctation of the aorta the systolic pressure in the arms does not have to be significantly higher than the average in an absolute sense The systolic pressure of one arm (usually the right) may be 20 to 30 mm higher than that of the other arm without necessarily implying that an added anomaly of one of the subclavian arteries is present In instances in which the difference

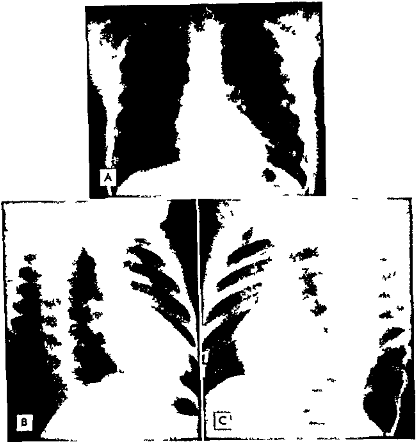


Figure 227 Posteroanterior and oblique radiograms of patient with moderately severe coarctation of the aorta Note the small aortic knob and evidences of left ventricular enlargement

in pressure between the two arms is more than 30 mm. of mercury some added abnormality such as stenosis or atresia of one of the subclavians must be assumed The pulsations of the abdominal aorta are often absent The feet may feel cooler to touch than the hands

At auscultation the heart sounds are usually normal in intensity and splitting although the second sound is often louder at the aortic area than at the second left interspace A systolic murmur is present in at least 75 per cent of the cases A stenotic systolic murmur, originating either at the aortic valve or the coarcted segment itself, may be heard at the second right interspace the suprasternal notch or, most characteristically over the back, at either side of the vertebral column A blowing systolic murmur at the lower left sternal border may also be heard but its origin has never been clear to me A continuous murmur (Fig 226) presumably originating from the extensive collaterals is often



Figure 228 Outline of the coarctation in conventional radiogram *A* E sign on the barium filled esophagus *B* The 3 sign of the coarctation in the posteroanterior view For explanation see text

heard in older children over the back at the second right interspace or even at the pulmonary area Early diastolic murmurs representing aortic regurgitation usually are attributed to the presence of a bicuspid aortic valve An apical mid diastolic rumble may be heard in children who have moderately severe cardiac enlargement but no additional intra cardiac anomaly

The roentgenologic picture may be entirely normal in a few cases In most instances however left ventricular enlargement is easily recog nized (Fig 227) Left atrial prominence is present in severe cases The main pulmonary artery and pulmonary vasculature are normal in size and activity The aorta is often hypoplastic and the impression of the descending arch on the barium filled esophagus often can be recognized by its E shape (Fig 228) The first arc of the E is the impression of that part of the aorta which precedes the coarctation the middle segment is the coarctation itself and the second arc is caused by the poststenotic dilatation In an overpenetrated film the aorta itself may be seen assum ing a big three shape consisting of the same components as the E only in the reverse "Scalloping" of the ribs due to erosion of the lower surface of the ribs by the collaterals is rarely seen in patients younger than eight years of age (Fig 30 p 40) At fluoroscopy the powerfully contracting left ventricle may be noted

The electrocardiographic pattern may vary from a normal tracing to one showing severe left ventricular hypertrophy depending on the severity of the coarctation (Fig 229) In our experience pure right ventricular hypertrophy is never present in patients with uncomplicated coarctation of the aorta after the age of six months

Cardiac catheterization (Fig 230) is seldom performed now in patients with simple coarctation The meager data available offer the

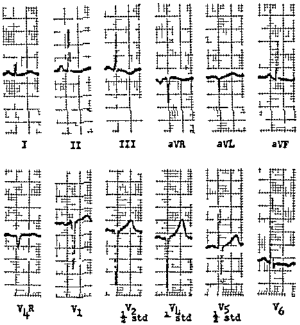


Figure 2_9 Electrocardiogram of patient with coarctation of the aorta Note definite evidences of left ventricular hypertrophy with T wave changes

following information [33, 3] (1) No left to right or right to left shunt exists (2) Pulmonary arterial and right ventricular pressures are normal or slightly elevated (3) Pulmonary capillary" pressure is normal or slightly increased The right brachial arterial systolic pressure is higher than the femoral arterial pressure and its peak is earlier (4) There is full arterial saturation (5) Systemic and pulmonary flows are equal Systemic resistance is markedly elevated pulmonary resistance is usually slightly higher than normal

Angiography is not the best method for studying patients with coarctation of the aorta On the other hand aortography with injection through the right brachial or the carotid artery is well suited to outline the site and length of the coarcted segment together with other vascular anomalies (Fig 122 p 125)

Course and Prognosis

The natural history of patients with simple coarctation of the aorta is rather uneventful at least through childhood If the dangers of bacterial endocarditis and rheumatic fever can be avoided by the judicious use of chemotherapy and chemoprophylaxis these patients seldom have any serious difficulty during the first two decades The only complications I have seen in this age group related to hypertensive encephalopathy or cerebrovascular accidents These are obviously severe even

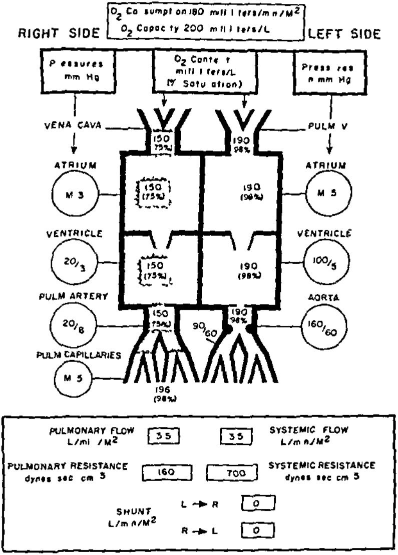


Figure 2_0 Diagrammatic presentation of cardiac catheterization data in patient with simple coarctation of the aorta

fatal complications but certainly they occur in much less than 5 per cent of the cases

According to the data collected by Reifenstein and his co workers [8] at postmortem examinations only a quarter of these patients can be expected to live through the fourth decade unless an operation is per formed The others die considerably earlier as a result of bacterial endocarditis ruptured aorta or hypertensive cardiovascular disease

Differential Diagnosis

The differential diagnosis of simple coarctation of the aorta should present no problem to the physician who as part of his routine physical

examination palpates the femoral pulses and tries to compare the pressures in the upper and lower extremities in patients with weak or late femoral pulses

Treatment

The treatment of coarctation of the aorta is surgical There are cogent reasons to suggest that all patients with coarctation of the aorta should be operated on to assure normal life expectancy and to eliminate the threat of bacterial endocarditis [219]

Since the initial work by Crafoord and Gross hundreds of patients with coarctation of the aorta have been treated surgically [87, 149] The optimal age of operation is between ten and fifteen years the operative risk in asymptomatic patients of this age group is less than 5 per cent However operation can be performed in patients of practically all ages although the mortality rate may be higher

The operation consists in division of the ligamentum arteriosum and wide excision of the coarcted segment Preferably the two aortic segments are then sutured directly together However if the gap left by the excision is too large an aortic graft may be inserted with success [149]

In 90 per cent of the patients the blood pressure falls to normal levels after this procedure and the femoral pulses can be easily palpated It is interesting to note that the systemic systolic pressure drops significantly within the first postoperative day but seldom returns to normal levels for two to three weeks after operation This is a good demonstration of the role of extra aortic (presumably peripheral arteriolar) factors in the maintenance of the hypertension of patients with coarctation of the aorta

As a rule at The Childrens Medical Center in Boston cardiac catheterization or aortography is not performed before operation in cases of simple coarctation of the aorta The clinical profile is usually outlined clearly enough to make these procedures unnecessary in the ordinary case The use of aortic grafts and the availability of a vessel bank make the accurate preoperative outline of the obstruction less important to the surgeon than it was formerly

Of course if the clinical picture is not a typical one all available means of investigation should be pursued before proceeding with an operation

Summary

Patients with a systolic pressure which is significantly higher over the arms than over the legs have coarctation of the aorta There is no evidence of a shunt in either direction Both the electrocardiogram and the roentgenogram show evidences of left ventricular hypertrophy Surgery is recommended for patients between ten and fifteen years of age

Coarctation of the Aorta with Congestive Failure in Infancy

Whereas the vast majority of patients with uncomplicated coarctation of the aorta associated with systemic left ventricle are asymptomatic throughout childhood in a minority of these infants (certainly less than 10 per cent) severe congestive failure develops Why these infants behave *so differently from others with similar anomalies is not known with* certainty Hypotheses based on the site of the ductal opening in the fetus and on the consequent differences in the development of collaterals have been proposed repeatedly[19, 370] to explain the difference between the clinical course of the two groups For reasons given elsewhere[199] I am unable to accept these hypotheses as the sole or even dominant ones rather I believe that the infants in whom congestive failure develops have the severest degree of coarctation at birth whereas in the others it develops progressively through the years

Clinical Picture

The description that follows is based principally on nine patients we have observed during the past ten years at The Children's Medical Center in Boston[199] In these infants symptoms usually begin within the first three months often the first few weeks of life Their weight gain is poor they are irritable and they have feeding problems dyspnea excessive perspiration and even occasional cyanosis

At physical examination these babies look sick with tachypnea tachycardia and perioral cyanosis As a rule the liver is enlarged, and moist rales may be heard over the lungs Peripheral edema is rare On auscultation a gallop rhythm is frequently noted and a systolic murmur is heard at the left sternal border in almost all instances The femoral pulses are weak and late in comparison with the right radial pulse or may be completely absent "Flush" pressure over the arms is at least 20 mm of mercury higher than over the legs the systolic pressure over the lower extremities however is rarely less than 50 mm of mercury

The roentgenologic picture is dominated by the features of congestive failure Cardiomegaly is maximal (Fig 231) the heart is globular in shape the beat is poor and there is passive congestion of the lungs

Electrocardiograms are always abnormal an abnormal degree of either right or left ventricular hypertrophy with or without changes in the T wave is present

Unfortunately no cardiac catheterization data or angiographic pictures are available for these patients Their precarious state on admission and their excellent response to treatment make most cautious observers shy away from these potentially dangerous investigative procedures

Course and Prognosis

If treated properly the congestive failure in these desperately sick infants disappears within a few days or weeks However a satisfactory

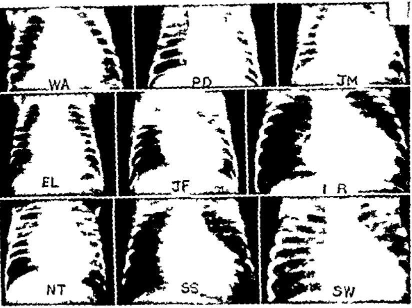


Figure 231 Posteroanterior radiograms of nine patients who went into congestive failure in infancy with coarctation of the aorta[199]

weight gain is not observed for many weeks or even months Eventually the infants start behaving normally and show no evidences of heart disease except the persistence of the murmur and the relative hyper tension of the upper extremities The heart size diminishes slowly but as the chest diameter grows it does assume normal size within a few years (Fig 232) The electrocardiographic changes are striking eventu ally all patients have left ventricular hypertrophy regardless of whether they exhibited left right or combined ventricular hypertrophy in early infancy (less than six months of age) (Fig 233) On the basis of our present rather limited experience we at The Children s Medical Center believe that if the pattern of left ventricular hypertrophy is not present by six months of age the patient probably does not have an uncompli cated coarctation of the aorta

Differential Diagnosis

As mentioned before the differential diagnosis should not present any particular problem if one remembers to feel for the femoral pulses of every patient and to measure both arm and leg pressures in all patients with weak or late femoral pulses

The condition most commonly considered in the differential diag nosis of these youngsters is primary myocardial disease (see p 209) Patients in both groups have in common severe cardiac enlargement without characteristic murmur congestive failure in early infancy and electrocardiographic evidences of left ventricular hypertrophy Once

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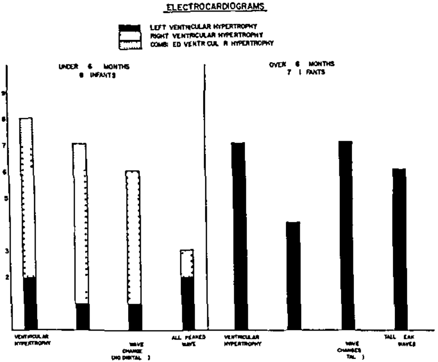


Figure 233 Electrocardiographic patterns in infants with coarctation of the aorta and congestive failure A comparison of tracings under six months and over six months of age[199]

SUMMARY

In infants with the severest degrees of aortic block congestive failure may develop The diagnosis is based on the presence of relative hypertension of the arms The electrocardiograms and roentgenograms are always grossly abnormal Anticongestive measures are usually successful in tiding these patients over until operation can be easily performed at an appropriate age

Coarctation of the Aorta with Left to Right Shunt

Whereas most patients with coarctation of the aorta have no communication between the systemic and pulmonary circuits and the systolic overload is carried entirely by the left ventricle a number of patients have been encountered in our clinic in whom coarctation of the aorta was accompanied by definite and even marked pulmonary plethora and right ventricular hypertrophy The anatomic situation usually reveals a ventricular defect or a patent ductus arteriosus opening above at or even below the site of the aortic block

CLINICAL PICTURE

Symptoms of exertional dyspnea fatigue repeated severe respiratory infections and failure to thrive manifest themselves early in these patients In terms of severity of symptoms these children rank between the patients with simple coarctation and the infants who have congestive failure On the whole the presenting complaints are more characteristic of patients who have left to right shunts than of those with coarctation of the aorta

At physical examination the patients are usually poorly developed Cyanosis and clubbing are absent but the fingers and toes are commonly fiery red Left chest deformity with a hyperdynamic combined ventricular impulse is the rule a thrill is frequently present at the left sternal border Evidences of mild left sided or right sided congestive failure may be noted Relative hypertension of the upper extremities is present

The observations at auscultation vary depending on the site of the communication between the systemic and pulmonary circulations We have not encountered any patient with an atrial communication so far If the communication is through a large patent ductus arteriosus (which incidentally is the most frequent site) and the pulmonary vascular resistance is relatively low a typical machinery murmur may be heard If the ductus is small or the pulmonary vascular resistance is near the systemic level only a second left interspace systolic murmur is noted If a ventricular defect forms the communication between the left and right sides the murmur typical of ventricular defect may be heard if the shunt is sizable If the opening is small little if any murmur can be observed The second sound at the pulmonary area is always booming a mid diastolic rumble is present if the shunt is large

The roentgenologic picture is characteristic of the large left to right shunt group (Fig 234) A significant degree of cardiac enlargement involving both ventricles and often the left atrium is present Pulmonary plethora with expansile pulsations is uniformly noted The main pulmonary artery is usually large and active the aorta varies considerably in size and activity

Electrocardiographically combined ventricular hypertrophy is noted without selective atrial hypertrophy Intraventricular conduction disturbance is common (Fig 235)

Cardiac catheterization (Fig 236) reveals the following (1) A left to right shunt is present at the ventricular or pulmonary arterial level (2) Pulmonary arterial and right ventricular hypertension exists (3) "Pulmonary capillary" pressure is elevated and the brachial arterial tracing is characteristic of coarctation of the aorta (4) There is arterial oxygen saturation of 91 to 95 per cent at rest without any significant difference in arterial oxygen content between the right brachial and femoral arteries (5) The pulmonary and systemic resistances are in

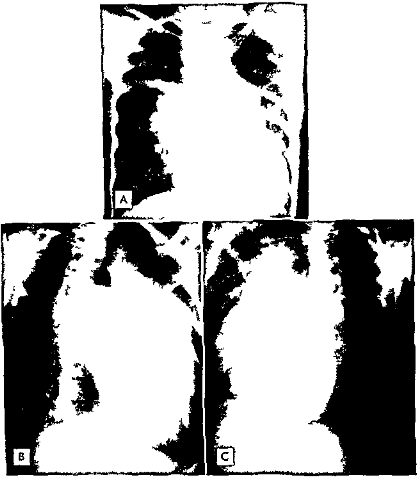


Figure 234 Posteroanterior and oblique radiograms of patient with coarctation of the aorta patent ductus arteriosus and a left to-right shunt

creased to varying degrees (6) Occasionally the catheter enters the aorta through a ductus or the left ventricle through a ventricular defect

As a rule angiocardiography fails to demonstrate the coarctation because of the dilution of the dye by the admixture of systemic blood Since the shunt is predominantly left to right the site of the communication is also seldom revealed

Course and Prognosis

The natural history of these patients is similar to that of others who have pulmonary hypertension associated with a communication between the two circuits The symptoms may be rather serious during infancy and early childhood Improvement perhaps secondary to the development of medial and intimal changes in the pulmonary arterioles occurs

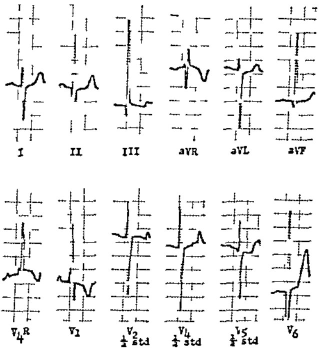


Figure 235 Electrocardiogram of patient with coarctation of the aorta and patent ductus arteriosus and a left-to-right shunt Note right axis deviation and combined ventricular hypertrophy

at about four to five years of age and lasts through the first two decades of life In early adulthood severe dyspnea on exertion develops this is followed by cyanosis congestive failure and death

Differential Diagnosis

The differential diagnosis of this syndrome is particularly important in respect to coarctation of the aorta with a systemic right and left ventricle that is a ductus carrying *pulmonary* arterial *blood* into the *distal* part of the *aorta* The similarities between the two groups are striking Patients in both groups have the typical blood pressure findings of coarctation of the aorta associated with some evidence of right ventricular hypertrophy as shown on the roentgenogram and the electrocardiogram in addition to pulmonary plethora Identification of the patients with systemic right and left ventricle is often possible by (1) definite evidence of cyanosis particularly over the left arm and the lower half of the body (2) severe or exclusive right ventricular hypertrophy according to the electrocardiogram and (3) absence of a significant or typical murmur At cardiac catheterization no evidence of a left to right shunt is found and the pulmonary arterial pressure is higher or identical with the pressure in the femoral artery The catheter often enters the descending aorta

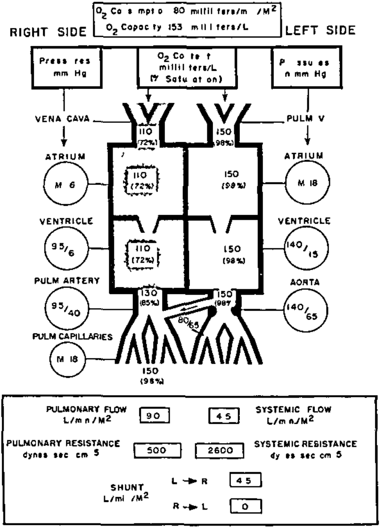


Figure _36 Diagrammatic presentation of findings of cardiac catheterization in a patient with coarctation of the aorta patent ductus arteriosus and left to right shunt

Treatment

Medical management of infections and congestive failure preoperatively and postoperatively obviously is indicated Surgical removal of the aortic block should be undertaken only if the parents are willing to assume the considerable risk (at least 25 per cent) inherent in the procedure At the time of operation the ductus arteriosus should be divided as well The purpose of surgical intervention over and above the usual ones listed in the section on simple coarctation of the aorta is to forestall progression of the pulmonary vascular disease In my

experience the more severe the pulmonary vascular obstruction the more hazardous is the operation If the pulmonary vascular resistance has reached a level at which the shunt is exclusively or predominantly from right to left then the operative risk becomes prohibitive as long as at least part of the time the shunt is from left to right operation should probably be attempted

Summary

The presence of right ventricular hypertrophy and pulmonary plethora in patients with *coarctation of the aorta suggests pulmonary* arterial hypertension and a communication between the systemic and pulmonary circuits The most common site of such a communication is a patent ductus arteriosus although ventricular defects are also seen occasionally Surgery is indicated as long as the shunt is predominantly left to right although the mortality rate is high

Coarctation of the Aorta with Systemic Right and Left Ventricles

Incidence

This type of congenital heart disease is relatively rare in patients *beyond the neonatal period It is a common cause of death in early* infancy however [60]

Anatomy

The principal circulatory abnormality in these patients is the presence of a large patent ductus arteriosus opening *below a coarctation* and *supplying the descending aorta with pulmonary arterial blood* The *coarcted segment may be short or may extend for several centimeters* along the aortic arch but is usually proximal to the origin of the left subclavian artery More often than not this is a complex anomaly it may be associated with congenital aortic or mitral valvular disease within the framework of endocardial fibroelastosis, but it also may occur in conjunction with ventricular septal defects and particularly with complete transposition of the great arteries I believe that this is the anomaly formerly referred to rather loosely as "infantile" coarctation of the aorta

Physiology

The physiologic effects of the malformation depend to a great extent on whether or not it is associated with other major cardiac anomalies If endocardial fibroelastosis or transposition of the great arteries is present obviously there will be severe and early alterations in the hemodynamic picture If coarctation of the aorta is the only anomaly present then during fetal life the circulation will not deviate significantly from normal except for the probable presence of left ventricular hypertrophy difficulties will arise however as soon as the shift toward extrauterine

circulation begins At this time the ductus arteriosus does not normally close but rather furnishes the lower half of the body with blood Thus right ventricular hypertension is maintained and the lower extremities receive unoxygenated blood

CLINICAL PICTURE

In the neonatal period there is severe left sided and right sided congestive failure dyspnea cyanosis and feeding difficulties *dominate* the picture At physical examination the cyanosis is usually generalized Occasionally a differential cyanosis is noted *the upper half of the body* especially the right hand being pink and the lower extremities showing definite blueness Chest deformity is common most often it consists in a significant increase in the anteroposterior diameter of the chest The cardiac impulse is usually hyperdynamic and is maximal at the xiphoid process

Auscultation seldom contributes anything specific to the diagnosis A blowing systolic murmur and possibly a low frequency diastolic murmur is usually heard at the lower left sternal border with the systolic component transmitting well to the neck and the back The second sound at the pulmonary area is usually accentuated Rales are commonly heard over both lung fields and the liver is enlarged but not pulsating

The *femoral pulses are weak and late, but seldom absent* In the majority of instances the systolic pressure in the upper extremities is definitely higher than in the legs however *there are infants with coarctation of the aorta and systemic right and left ventricle in whom no pressure gradient between the upper and lower extremities exists because of the simultaneous presence of a large ventricular defect resulting in equal systolic pressure in the two circuits*

Roentgenologic examination reveals marked cardiac enlargement involving all chambers with pulmonary vascular engorgement (Fig 237) At fluoroscopy the cardiac beat is poor and expansile pulsations are rarely noticeable None of the characteristic evidences of coarctation of the aorta on the esophagus mentioned before can be noted in these babies

The electrocardiogram invariably reveals right ventricular hypertrophy of abnormal degree with or without simultaneous evidences of left ventricular hypertrophy (Fig 238)

No adequate angiocardiographic or cardiac catheterization studies on our patients are available they are usually too sick to be subjected to these procedures The one physiologic evidence which is relatively easy to obtain and furnishes a definite clue to the diagnosis is the measurement of the pressure and the oxygen content simultaneously in the right brachial and one of the femoral arteries As in cases of a "reversed ductus" the oxygen content in the femoral artery is appreciably lower



Figure 237 Posteroanterior and oblique radiograms of patient with coarctation of the aorta and systemic left and right ventricles This patient was about nine years old when this film was taken

than that in the right brachial artery but in addition in most instances the systolic pressure in the lower extremities is also significantly lower than it is in the radial artery

Course and Prognosis

Most of these babies die within the first days or weeks of life in congestive failure A minority of them perhaps through adjustments in the pulmonary vasculature [106] manage to survive the neonatal period and may live for several years The activities of these patients are always severely limited they are markedly cyanotic and are bordering on con

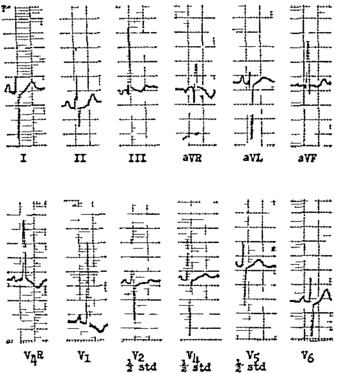


Figure 238 Electrocardiogram of patient with coarctation of the aorta and systemic left and right ventricles Note the marked degree of right ventricular hypertrophy

gestive failure most of the time Death usually occurs within the first or second decade

Differential Diagnosis

The differential diagnosis of coarctation of the aorta with systemic right and left ventricle may be approached from two aspects (1) The differentiation of this specific type of coarctation of the aorta from all other types mentioned may be accomplished with certainty only if femoral arterial and right brachial arterial pressures and oxygen contents are measured simultaneously The anomaly should be suspected however in any patient more than six months old with coarctation of the aorta in whom right ventricular hypertrophy is present in the electrocardiogram

2 The second aspect of the differential diagnosis is the problem of newborn infants in severe congestive failure who may or may not have coarctation of the aorta Obviously the discovery of relative hypertension in the arms establishes the diagnosis easily however sometimes in newborn infants it is difficult to determine the blood pressure and to interpret it correctly As a rule if some degree of cardiac compensation can be accomplished with vigorous anticongestive measures the correct

blood pressure values are much easier to determine If compensation cannot be achieved and an accurate diagnosis is desired for these desperately sick infants cardiac catheterization may be attempted more to rule out other diagnoses than to establish the presence of a coarctation of the aorta

Treatment

Medical treatment may improve congestive failure to a considerable extent but obviously it cannot postpone for many years the inevitable end

In older children operation has almost always been fatal the situation being identical to that in patent ductus arteriosus with pulmonary vascular obstruction [7, 8] In some of the younger infants in whom presumably permanent organic pulmonary vascular obstruction has not yet developed surgery has been successful [31, 370]

At The Children's Medical Center in Boston our policy is to treat all infants with coarctation of the aorta and congestive failure with anticongestive measures If prompt and lasting improvement occurs, we assume that the patient has coarctation of the aorta with systemic left ventricle and congestive failure If the medical regimen is unsuccessful operation at high risk *is recommended and the surgeon is warned of* the possibility that a ductus arteriosus may open below the coarctation and may carry blood from the right to the left side of the circulation

Summary

Coarctation of the aorta with systemic right and left ventricle should be suspected in every patient in whom there is relative hypertension of the upper extremities and electrocardiographic evidence of right ventricular hypertrophy in infants less than six months of age this represents only presumptive evidence but in older children it is highly suggestive The diagnosis may be confirmed by simultaneous right brachial and femoral arterial blood samples and pressure tracings Medical management is usually not successful At best it may prolong life for only a few years In older patients surgery is hopeless in young infants good results have been reported in a few instances

AORTIC STENOSIS

General Principles

Until recently aortic stenosis was exclusively an anatomic concept a narrowing of the left ventricular outflow tract at postmortem examination was considered to indicate aortic obstruction The clinical profile was based entirely on patients whose diagnosis was confirmed anatomically a situation similar to that of pulmonary stenosis ten years ago that is before the introduction of right heart catheterization With

Table 30

DIFFERENTIAL DIAGNOSIS OF VARIOUS TYPES OF COARCTATION OF THE AORTA

	CLINICAL PICTURE													PHYSIOLOGY					COURSE & REMARKS
		M			X-RAY					EK				SHUNT		PRESSURE			
	C	SYST	CON	DIAST	AE	RVE	VE		PV	P	VH	VH	AH	L-R	R-L			AO	
SIMPLE COARCTATION OF THE AORTA		STENOTIC MECHANICAL [illegible] II–III [illegible]		OCC RUMBLE OR EARLY BLOW						*		+						HIGHER THAN	[illegible] COURSE THROUGH CHILDHOOD, DEATH IN MIDDLE AGE FROM [illegible]
COARCTATION OF THE AORTA C CF IN INFANCY		G III PRECORDIAL		OCC RUMBLE					PASSIVE CONGESTION									AS ABOVE	[illegible] DEVELOPE IN INFANTS OF UNDER 4 MO. USUALLY RESPOND WELL TO ANTICONGESTIVE MEASURES
COARCTATION OF THE AORTA C L→R SHUNT		G II [illegible] BACK		RUMBLE OR EARLY BLOW					++	–		+					+	AS ABOVE BUT SHOW [illegible]	STORMY INFANCY, RELATIVELY STABLE CHILDHOOD, DEATH IN EARLY ADULTHOOD [illegible] OF SHUNT MOST LIKELY PDA OR VSD
COARCTATION OF THE AORTA C SYSTEMIC R-L	++	G II AOR SPECIFIC				++				–	++	–						HIGHER THAN	MOST OF THEM DIE IN EARLY INFANCY, ARE MORE CYANOTIC [illegible] FEW SURVIVE FIRST DECADE

See Table 28 (p 303) for explanation of symbols

the advent of left heart catheterization it seems likely that a physiologic definition i e presence of a gradient between the left ventricle and the aorta will supplant the old anatomic concept, and new ideas in terms of frequency of the condition clinical picture and prognosis will be introduced For the purposes of this discussion however the diagnostic criteria based on the old anatomic definition of aortic stenosis will have to prevail Therefore at present, the diagnosis of aortic stenosis is made on the basis of the following clinical findings (1) a second right inter space "stenotic" systolic murmur with good transmission to the neck and the back, (2) evidences of left ventricular hypertrophy or enlargement on the roentgenogram or electrocardiogram and (3) no evidence of a left to right or right to-left shunt

A number of other clinical findings will be mentioned later but these three are the only ones essential for the diagnosis

According to the latest data concerning adults with aortic stenosis [44] the principal etiologic factors causing this anomaly are rheumatic fever and arteriosclerosis Since for all practical purposes arteriosclerosis does not occur in childhood and since the development of rheumatic aortic stenosis in all well documented cases takes several years presumably all young children with aortic stenosis have a congenital lesion One further point, first emphasized by Kiloh[189] and later mentioned by Levine [44] is that a history of rheumatic fever is rare in pure aortic stenosis whereas it is common in patients with combined aortic or multi valvular lesions On this basis then all children in whom the murmur of aortic stenosis is discovered before the fourth year of age certainly have a congenital lesion Most likely all those less than eight years old in whom such a defect is diagnosed and who have no history of rheu matic fever were born with the disease A third category of patients (not

to be discussed here) are adults with pure aortic stenosis in whom the presence of a congenital lesion cannot be proved but should be suspected[169]

Incidence

Congenital aortic stenosis thus defined is not one of the more common congenital cardiac anomalies but it is far from being a rare condition No accurate figures on its frequency in living patients are available Abbott found twenty three instances in 1000 postmortem examinations We have had sixty seven patients with congenital aortic stenosis in our clinic within the past five years Taking several factors into consideration it could be estimated that more than 1 per cent of the children two years old or older with congenital heart disease have pure aortic stenosis

Anatomy

Congenital aortic stenosis may be divided anatomically into valvular and subvalvular types In the valvular type the cusps are thickened and may be fused to form a dome shaped diaphragm with a central opening and a poststenotic dilatation of the ascending arch of the aorta Whether congenital valvular stenosis is truly a developmental anomaly or is the result of endocardial fibroelastosis localized at the aortic valve is difficult to decide clinically but both situations probably exist Subvalvular or subaortic stenosis usually consists of a fibrous ring encircling the left ventricular outflow tract approximately 5 to 10 mm below the aortic ring and often involving the anterior cusp of the mitral valve This defect in contrast to the valvular lesion is probably always developmental in origin

Embryologically speaking the valvular lesion probably occurs at about the seventh intrauterine week and thus later than the subvalvular This may be the reason why valvular lesions are not commonly associated with septal defects whereas occasionally subaortic stenosis does occur with a ventricular defect Conversely since endocardial fibroelastosis commonly causes valvular aortic stenosis a combination of valvular aortic with mitral stenosis and even coarctation of the aorta may occur whereas an association of subaortic stenosis (presumably a developmental lesion) with coarctation is practically never seen

Physiology

The principal physiologic burden of aortic stenosis is carried by the left ventricle The problem is to maintain an adequate flow and pressure in the systemic circuit through an orifice which is reduced in size Animal experiments[3] indicate that a reduction of the lumen of the aorta to at least 25 per cent of its original size is necessary before significant interference with blood pressure pulse pressure or cardiac output occurs Since the calculated area of the aortic orifice in adults is usually esti

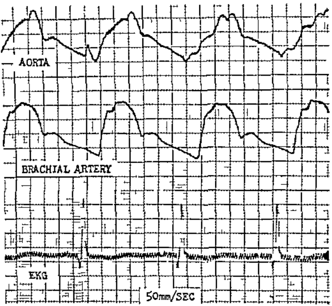


Figure 239 Simultaneous aortic and brachial arterial tracings with an electrocardiogram as a reference tracing in patient with severe aortic stenosis Note the slow rise of the tracing particularly in the aorta and the wide plateau with the double notch A dicrotic notch is present in both the aortic and brachial arterial tracings

mated to be 2 5 to 3 5 sq cm probably no significant interference with the circulation occurs at least in the acute experiment until it is reduced to less than 0 6 sq cm [144] If the size of the aortic orifice is reduced below the critical level compensatory mechanisms are called into action the left ventricular diastolic volume and considerably later the end diastolic pressure increase in order to elevate the left ventricular peak systolic pressure Isometric contraction is slightly longer ejection time and total systole become considerably prolonged The pressure within the left ventricle may rise as high as 200 to 250 mm of mercury but seldom higher The aortic and peripheral arterial tracings show a slow rise with a low anacrotic notch or slur a wide plateau with vibrations corresponding to the systolic murmur and a slow descent often with an absent dicrotic notch (Fig 239) As the left ventricle attempts to maintain the necessary systolic pressure it hypertrophies and its oxygen needs increase At the same time the coronary blood flow does not increase proportionately it may even become smaller because of (1) involvement of the coronary ostia in the stenotic process (2) shortening of diastole the time when the coronary arteries fill (3) the squeezing effect on the coronaries by the high left ventricular systolic pressure and (4) possibly a suction effect on the coronary ostia by the high velocity flow through the small aortic opening As a result of all these factors and the increased

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Figure 241 Phonocardiogram of patient with aortic stenosis Note the early diamond shaped murmur at the second right interspace In the left hand tracing the carotid artery serves as a reference whereas in the right hand tracing an electrocardiogram is used for reference tracing

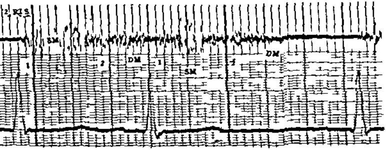


Figure 242 Phonocardiogram of a patient with aortic stenosis on a congenital basis associated with aortic regurgitation Note the early diamond shaped systolic murmur followed by the long diastolic murmur lasting throughout most of diastole

was maximal at the second right interspace in the others it was best felt at the lower left sternal border or even at the apex At auscultation the heart sounds seemed essentially normal usually the second sound in the aortic area was normal or slightly diminished only seldom accentuated. A late systolic click at the aortic area was noted in some patients [83] The systolic murmur at the aortic area was at least grade 3 to 4 in intensity and was always stenotic Careful auscultation suggests and phonocardiograms demonstrate that the diamond of the aortic stenosis murmur reaches its peak early at about the first third of systole (Fig 241) Swedish observers found good correlation between the intensity of the murmur its duration the time of its maximal intensity and the severity of the lesion [191] So far our experience has failed to confirm this observation

Diastolic murmurs were noted in twelve of our sixty seven patients Most of these were grade 2 to 3 early high frequency blows (Fig 242) some were mid-diastolic rumbles The systolic pressure over the upper and lower extremities was within normal limits in most instances The pulse pressure was normal in fifty four of the sixty seven patients in eight instances it was less than 20 and in five it was more than 50

Roentgenologic examination is not specific in most cases Slight left ventricular enlargement was noted in about half of our patients in

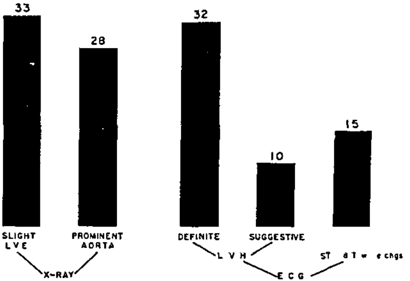


Figure 243 Summary of clinical laboratory tests in sixty seven patients with congenital aortic stenosis

twenty eight the ascending aorta was prominent and showed increased pulsations (Fig 243)

The electrocardiogram (Fig 243) was within normal limits in only 25 per cent of our cases in the others left ventricular hypertrophy of varying degree was observed Some of these completely asymptomatic patients showed the most severe progressive left ventricular hypertrophy with evidences of strain (Fig 244)

Right heart catheterization usually shows no significant abnormalities at rest However in the severest cases of aortic stenosis there is usually a relatively fixed cardiac output and a moderately severe increase in the "pulmonary capillary" pressure on exercise Left heart catheterization may furnish direct evidence in regard to the nature and severity of the lesion by enabling the cardiologist to actually measure the gradient across the aortic valve and by relating this to the cardiac output to determine the size of the aortic orifice Patients with the severest aortic stenosis have a gradient across the valve of 100 to 150 mm of mercury although probably any pressure difference between the left ventricle and the aorta of more than 40 mm of mercury indicates significant disease

Analysis of the brachial arterial pressure tracing alone or within the framework of cardiac catheterization has been advocated by Goldberg and his associates[135] as the most practical means of assessing the severity of the stenosis These authors pay particular attention to the time interval between the onset of the rise in the brachial arterial tracing and the peak of systole in their patients with aortic stenosis this time interval averaged 0 20 second whereas in a few normal persons the average was 0 08 second Other features of the brachial arterial tracing

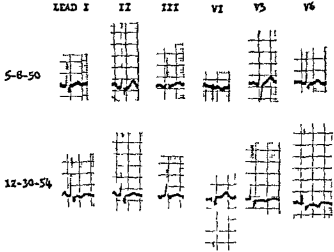


Figure 244 Severe left ventricular hypertrophy developing in a patient with congenital aortic stenosis in a four year period In the top tracing in 1950 T waves in leads I and V_6 are upright whereas in the bottom tracing in 1954 T waves in these leads are inverted Also note the increase in voltage in the chest leads in 1954

which they mentioned are the presence of an anacrotic notch and a double peaked plateau In our hands and in the experience of others the brachial arterial tracing although a useful measurement indeed has failed to give adequate information in all instances

Angiocardiography is of little help in the diagnosis or evaluation of aortic stenosis

Course and Prognosis

Although most children with aortic stenosis have few if any symptoms and lead entirely normal lives through childhood and adolescence we have had five sudden deaths among our sixty seven patients (Table 31) On the other hand we know from recent studies on a large number of patients with pure aortic stenosis that their average life expectancy is more than sixty years [44]

Therefore the question confronting the pediatric cardiologist is which patient will die of coronary disease and congestive failure at the age of sixty and which will drop dead suddenly in early adolescence after minimal physical exertion such as riding a bicycle? Obviously there is no simple answer The only clinical hint as to the severity of the disease is furnished by the electrocardiogram It is my opinion that although one cannot be certain that an aortic or subclavian aneurysm may not rupture in a child with a perfectly normal electrocardiogram the presence of severe and progressive left ventricular hypertrophy with strain increases significantly the danger of sudden death

Death from aortic stenosis in children is almost never due to con

*Table 31 Causes of Sudden Death in 5 Patients among 67 Children with Congenital Aortic Stenosis**

Age	Activity at Death	Symptoms	Electrocardiogram	Autopsy
5	Playing in yard	—	Normal	Rupture of L subclavian aneurysm valv stenosis
6	?	Fatigue	L V H without T changes	Not done
8	Playing in field	Fatigue, Dyspnea, Pallor } exertion	L V H with S T and T changes	Not done
13	Coming home from play	Dyspnea + +	L V H with S T and T changes	Not done, Extreme L V H
14	Bowling and basketball	? Asthma	L V H with T changes	Myocardial fibrosis, Valvular stenosis

* Note that in only one out of the 5 was the electrocardiogram normal In 4 of them severe left ventricular hypertrophy was present in 3 of these with marked S-T and T wave changes

gestive failure probably ischemic heart disease with ventricular fibrillation is the most common cause of death Death in congestive failure may occur in infants with severe aortic stenosis

Differential Diagnosis

The differential diagnosis of congenital aortic stenosis usually does not offer any great difficulties

Rheumatic Aortic Stenosis Usually rheumatic aortic stenosis can be excluded easily on the basis of the age of the patient the absence of aortic regurgitation or multivalvular involvement and the absence of a history of rheumatic fever

Aortic Regurgitation Pure or dominant aortic regurgitation of a congenital nature is extremely rare[354] although I believe I have seen patients with this anomaly Obviously the etiologic evidence in these cases is purely circumstantial The murmur and pulse pressure should make it easy to distinguish free aortic regurgitation from aortic stenosis associated with the slight murmur of aortic regurgitation

Pulmonary Stenosis In its mildest form pulmonary stenosis without right to left shunt may be difficult to differentiate from mild aortic stenosis Both conditions are characterized by a "stenotic" systolic murmur and in both electrocardiograms and roentgenograms are normal The two most useful clues in the differential diagnosis are the site of maximal intensity of the murmur and the relative intensity of the second sound in the aortic and pulmonary areas Occasionally no accurate decision can be made on the basis of these criteria however Under these circumstances cardiac catheterization may prove or disprove the presence of pulmonary stenosis However the more conservative cardiologist probably will do nothing about these children since whichever disease they

have is mild enough not to cause any significant change in the electro cardiogram and therefore the differential diagnosis is of no real prognostic or therapeutic importance

Small Ventricular Septal Defect The differences between aortic stenosis and a small ventricular septal defect of the Roger type are discussed on page 312

Treatment

Children with severe valvular aortic stenosis should be treated surgically by means of valvotomy [37] Whether the transventricular or the transaortic approach is to be preferred is a subject of lively controversy

The severity of the lesion can be roughly assayed by electrocardiography and more definitely appraised by left heart catheterization At this time I believe that the aortic valve area and the gradient across the aortic valve should be determined in every patient with congenital aortic stenosis in whom electrocardiograms show severe and progressive changes If the patient has a large enough left atrium (on x ray examination) to allow left atrial puncture combined left and right heart catheterization should be performed If the left atrium seems too small thoracotomy may be performed and the pressure gradient and the cardiac output may be determined at operation Probably valvotomy should be performed in every patient who has a valve area less than 0 6 sq cm or a pressure gradient across the valve of 40 mm of mercury or more

It is difficult if not impossible to determine clinically whether the stenosis is valvular or infundibular It is my distinct impression however that the most severe cases of stenosis that is those in which operation should be contemplated are usually valvular This impression is reinforced by data in the literature [37] and by personal communications from my colleagues [18 197 33]

The success of surgical treatment is difficult to evaluate Objectively the transaortic gradient may be reduced at the operating table (Fig 245) and the electrocardiogram usually looks better six to eight months after operation (Fig 246) The patients may feel better but many of them have no symptoms to begin with To further complicate the evaluation of surgical treatment, aortic regurgitation apparently develops post operatively in most patients (Fig 247) the influence of this on life expectancy is hard to determine

The operative mortality in children is low less than 10 per cent in the hands of capable surgeons

Summary

Children with pure aortic stenosis probably have congenital disease Whether the stenosis is valvular or subvalvular cannot be determined clinically with accuracy but the patients with the severest disease usually

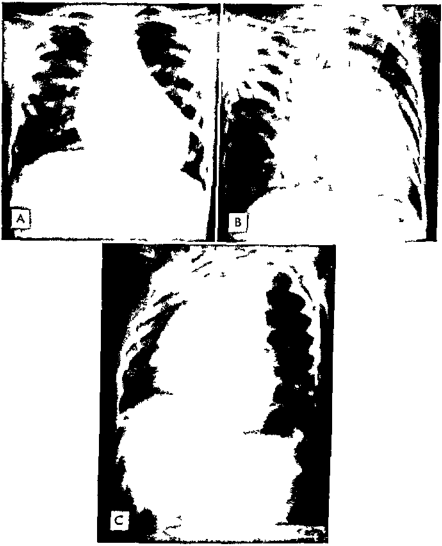


Figure 248 Posteroanterior and oblique radiograms of a patient with congenital aortic regurgitation

described in the section on rheumatic aortic regurgitation (p 162) A slight aortic systolic murmur was noted in all these patients but no physiologic consequences of aortic stenosis were observed No cardiac catheterization or angiographic data are available One patient has severe attacks of paroxysmal atrial tachycardia with the Wolff Parkinson White syndrome but otherwise all of them are essentially symptom free at the end of several years of observation (Figs 248 249)

These patients are mentioned to direct attention to the fact that congenital aortic regurgitation probably does exist although it must be an extremely rare condition

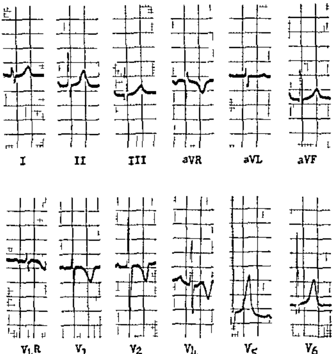


Figure 249 Electrocardiogram of a patient with congenital aortic regurgitation

VASCULAR RINGS

Vascular rings are congenital anomalies of the aortic arch and its tributaries causing varying degrees of compression of the trachea and the esophagus Strictly speaking since no cardiac dysfunction is con nected with them, these lesions probably should not be discussed in a textbook on pediatric cardiology However a brief discussion is included for the sake of completeness and because information concerning these anomalies is important to the pediatrician Most of the data that follow have been compiled by Doctors Robert E Gross[1, 1] and Edward B D Neuhauser[154] of The Children s Medical Center of Boston

Anatomy

Although if one reviews the complicated development of the aortic arch and the branchial artery, one can easily recognize that there are innumerable possible variations and permutations on normal development only those anomalies severe enough to cause symptoms of sufficient frequency to be of practical significance will be discussed here There are five such lesions (1) double aortic arch (Fig 250) (2) right aortic arch with a left ligamentum arteriosum (Fig 251) (3) anomalous right subclavian artery (Fig 252) (4) anomalous innominate artery (Fig 253) and (5) anomalous left common carotid artery (Fig 254) In all

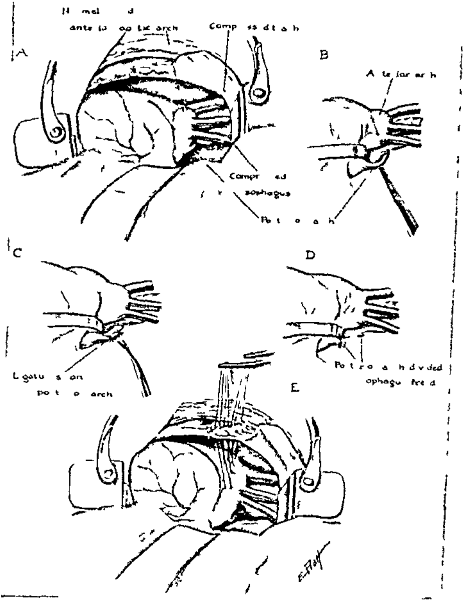


Figure 250 Double aortic arch with surgical division.[151]

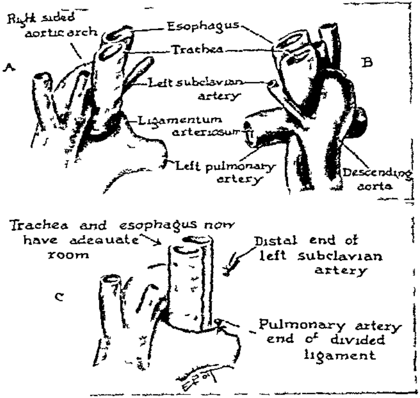


Figure 251 Right aortic arch with left ligamentum arteriosum and its surgical division [1]

these anomalies the trachea and the esophagus which normally lie behind the vascular structures are surrounded by vessels

INCIDENCE

These lesions are comparatively common Within ten years Gross[1 1] has seen fifty seven patients with vascular rings Their relative frequency in his series is as follows double aortic arch, twenty-one right aortic arch with left ligamentum arteriosum fifteen anomalous subclavian arteries twelve anomalous innominate arteries seven anomalous left common carotid artery two

PHYSIOLOGY

The consequences of vascular rings can be measured in terms of encroachment on the trachea and the esophagus If the esophagus is principally affected, swallowing difficulties and vomiting occur whereas, if tracheal compression dominates wheezing, cyanosis and frequent episodes of pneumonia may result.

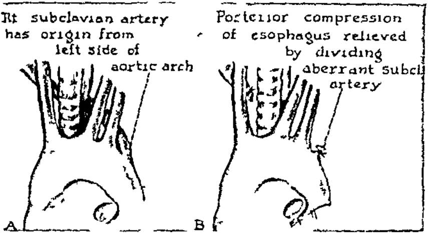


Figure 252 Anomalous right subclavian artery and its surgical division [151]

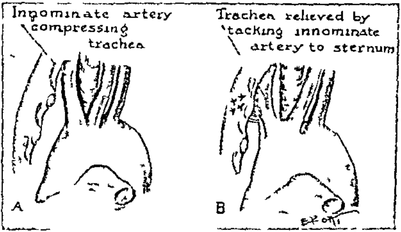


Figure 253 Anomalous innominate artery with its surgical division [151]

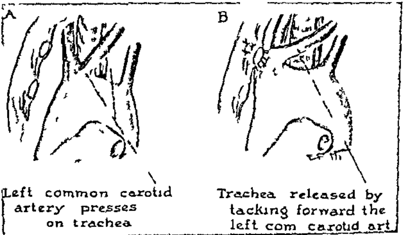


Figure 254 Anomalous left common carotid artery and its surgical division [151]



Figure 255 Esophagogram of patient with double aortic arch

Clinical Picture

The clinical picture varies considerably depending on the nature of the malformation and the severity of the obstruction Many of the malformations particularly those of the anomalous right subclavian artery cause no symptoms Others may cause severe difficulties even death in young infants

At physical examination the children with severe obstructions are extremely sick The typical patient is a small, poorly developed baby with a great deal of wheezing and stridor To reduce the tracheal compression to a minimum the child tends to keep his head in hyperextension Vomiting occurs frequently Feeding exaggerates the stridor and the wheezing and may cause cyanosis Many severe respiratory infections occur During these respiratory infections the stridor and vomiting increase as well

In some instances one is impressed by the fact that although these infants are hungry and start to eat eagerly after a few mouthfuls they cannot swallow and may even regurgitate In others small quantities may be taken easily whereas large amounts cause discomfort and vomiting.

The heart on physical examination seems completely normal the lungs may or may not show evidences of pneumonia Mild abdominal distention is commonly present

The electrocardiograms are well within normal limits

Roentgenograms of the heart and cardiac fluoroscopy show no significant abnormalities However by careful observation of the passage of barium along the esophagus it is possible not only to establish the diagnosis of a vascular ring but also often to identify the structures

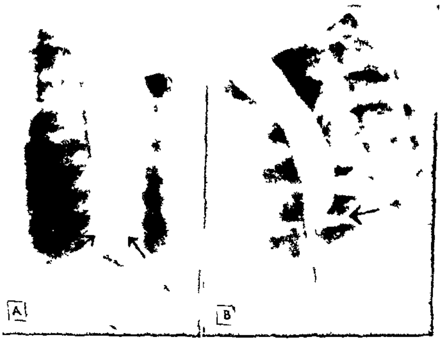


Figure 256 Esophagogram of patient with right aortic arch and left ligamentum arteriosum



Figure 257 Esophagogram of patient with anomalous right subclavian artery

making up this constricting ring Figures 255 256 and 257 demonstrate the impression of the compressing vessels on the barium filled esophagus In order to ascertain the degree of compression on the trachea as well a Lipiodol tracheogram should be obtained after the diagnosis of a vascular ring has been established by the history physical examination

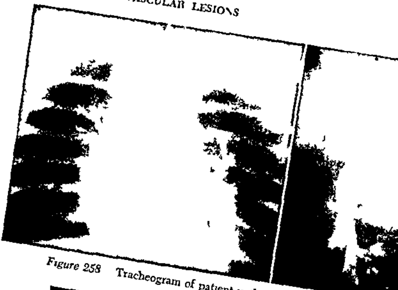


Figure 258 Tracheogram of patient with double aortic arch

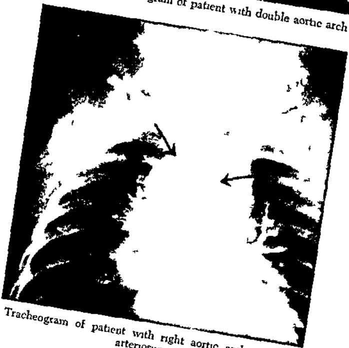


Figure 259 Tracheogram of patient with right aortic arch and left ligamentum arteriosum

and roentgenogram of the barium filled esophagus A combined roentgenologic examination of this sort almost always reveals the entire nature of the vascular anomaly Figures 258 259 and 260 are tracheograms of the three most common lesions

Cardiac catheterization has not been performed in any of our patients with this syndrome



Figure 260 Tracheogram of patient with anomalous innominate artery Note the simultaneous filling of the esophagus and the trachea

Angiocardiograms are seldom necessary the diagnosis is usually clear from the roentgenologic studies of the barium filled esophagus and the Lipiodol filled trachea In the few instances in which angiocardiography was performed the vascular structure was distinctly outlined

Course and Prognosis

The symptoms of patients with vascular ring depend to a great extent on the nature of the anomaly and the severity of the compression Children with double aortic arch usually have symptoms consisting principally of respiratory embarrassment early in infancy However a few infants are practically symptom free As a rule patients with a right aortic arch and left ligamentum arteriosum do not have symptoms until later in childhood the compression here also affects primarily the trachea but it is not as severe as in patients with double aortic arch In contrast to these two anomalies the aberrant subclavian artery seldom causes any respiratory distress The majority of the subclavian anomalies are completely asymptomatic and are compatible with a normal life expectancy However in severe cases dysphagia may result

The effects of anomalous innominate or left common carotid arteries are primarily respiratory and usually occur early in infancy

Differential Diagnosis

Primarily the differential diagnosis of vascular rings concerns con ditions causing repeated chronic respiratory infections in infants and children Among these pancreatic fibrosis and tracheo esophageal fistula without esophageal atresia should be considered and excluded by ap propriate clinical chemical and roentgenologic examinations

Treatment

If the vascular rings cause symptoms and if there are radiologic evidences of compression operation should be performed The appro priate surgical techniques have been described in detail [151] The aim of the surgical approach is to relieve the obstruction in cases of double aortic arch right aortic arch with left ligamentum arteriosum and anomalous right subclavian artery this is accomplished by severing the appropriate vessel In cases of anomalous innominate or left common carotid arteries obviously this cannot be accomplished without causing severe damage to the brain Therefore in these instances the compres sion is relieved by simply pulling the vessels and fastening them an teriorly and to the side

Summary

Anomalies of the aortic arch may cause compression of the esoph agus and the trachea Depending on the nature of the anomaly and the site of maximal compression respiratory symptoms or dysphagia may dominate the picture Particularly characteristic is the appearance of respiratory symptoms at feeding time After preliminary barium studies of the esophagus and Lipiodol examination of the trachea appropriate surgical repair is recommended if the symptoms are real and the com pression is demonstrable

UNDERDEVELOPED LEFT VENTRICLE WITH MITRAL AND AORTIC STENOSIS OR ATRESIA

Incidence

This is an extremely rare and complex anomaly Abbott[1] encoun tered seventeen such cases and Rossi [93] mentioned thirty six instances described in the literature We have seen fourteen patients with this condition in the past ten years at The Children s Medical Center in Bos ton Eight of our fourteen patients were males

Anatomy

The lesion may be developmental in origin in which case it is asso ciated with other congenital cardiac abnormalities (such as ventricular septal defect) or it may occur with endocardial fibroelastosis without accompanying malformations

Anatomically the most important finding is the hypoplastic or absent left ventricle with both the mitral and the aortic orifices being small or even completely absent In addition the aortic arch is narrow up to the point of entry of the ligamentum arteriosum Evidences of endocardial fibroelastosis may be plentiful within the wall of the small left ventricle the left atrium and the valves Whether fibroelastosis involving the left ventricle is the primary lesion leading secondarily to functional hypoplasia of the left ventricle or whether the hypoplasia of the ventricle is the basic defect is impossible to ascertain

Physiology

Functionally the heart acts like a two chambered heart The pulmonary venous blood enters the left atrium from which it is shunted through the foramen ovale into the right atrium Here it mixes completely with the caval blood and enters the right or single ventricle through the tricuspid valve Only a small amount if any of the right ventricular blood may enter the left ventricle and the aorta through a ventricular defect The bulk of the right ventricular output enters the pulmonary artery which furnishes blood to the pulmonary veins on the one hand and to the systemic circuit through an unusually large patent ductus arteriosus on the other This arrangement results in a complete mixing of the blood in the right atrium a poor systemic output and a relatively inadequate pulmonary blood flow

Clinical Picture

Some of the sickest babies with congenital heart disease are patients with this condition Dyspnea and cyanosis are usually noted immediately after birth or certainly within the first few days of life Feeding difficulties and poor weight gain are the rule almost without exception

On physical examination these infants are usually poorly developed and in a great deal of distress Their color is pale with deep cyanosis and their breathing is rapid and labored Evidences of left sided and right sided congestive failure are present Left chest deformity and marked cardiac enlargement may be noted within the first few hours or days of life As a matter of fact these babies may show a greater degree of cardiomegaly earlier in life than patients with any other cyanotic congenital defect

Auscultation reveals surprisingly little of significance The second sound at the pulmonary area may be accentuated often but not invariably a grade 3 systolic murmur is heard at the left sternal border However because of the critical condition of these babies no accurate or detailed auscultatory or phonocardiographic studies are available anywhere It is possible that as in so many other congenital cardiac anomalies further careful study of the heart sounds and murmurs may reveal a more consistent pattern than is apparent now

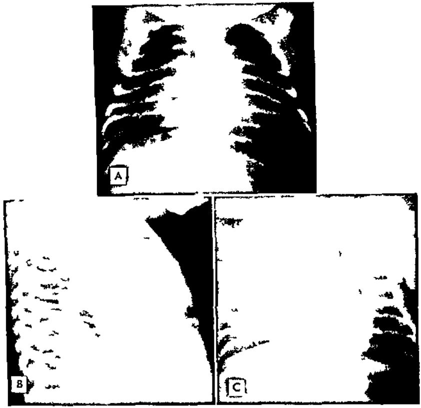


Figure 261 Posteroanterior and oblique radiograms in patient with congenital mitral and aortic atresia with hypoplastic left ventricle

Roentgenologically (Fig 261) the most impressive feature is the presence of marked cardiac enlargement with normal or increased pulmonary vasculature at an early age All cardiac chambers especially the right ventricle seem big and the pulsations under the fluoroscopic screen are usually feeble

The electrocardiogram shows marked right ventricular hypertrophy (Fig 262) sometimes with evidences of left ventricular hypertrophy

No cardiac catheterization or angiographic data are available on any of these critically ill babies

Course and Prognosis

The disease has a rapid downhill course death occurring within the first few days or weeks of life Rossi [93] stated that the longest survival on record was four months Death follows with evidences of cyanosis and congestive failure The longest survival in our group of patients was nineteen weeks

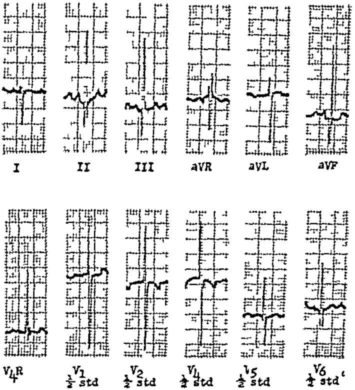


Figure 762 Electrocardiogram of patient with aortic and mitral atresia and hypoplastic left ventricle Note the presence of marked right ventricular hypertrophy in the chest leads and evidences of left atrial hypertrophy in leads V_5 and V_6

Differential Diagnosis

The differential diagnosis is a most difficult one since it involves all conditions giving rise to marked cardiac enlargement, cyanosis and congestive failure in young infants Among these transposition of the great arteries coarctation of the aorta with systemic right and left ventricle total anomalous pulmonary venous drainage with inadequate interatrial communication and all variations of the two chambered and three chambered hearts must be mentioned No clear cut clinical differentiation among these conditions is possible at present if and when technically and philosophically it is feasible to perform selective angiograms on all critically ill newborn infants an attempt at differential diagnosis may be possible

Treatment

Unfortunately there is no treatment available beyond the inadequate symptomatic measures designed to cope with infection and congestive failure

Summary

A patient who has an underdeveloped left ventricle with mitral and aortic stenosis or atresia is usually critically ill with an extremely large heart cyanosis and congestive failure The findings at auscultation are nonspecific the electrocardiogram shows right ventricular hypertrophy and roentgenograms reveal diffuse cardiac enlargement No treatment is available Death occurs within the first days or weeks of life

CONGENITAL MITRAL STENOSIS

In contrast to the patients with underdeveloped left ventricle and mitral and aortic atresia a functioning left ventricle is present in the anomaly designated as "congenital mitral stenosis" The dividing line between the two malformations cannot be drawn sharply in many instances the difference is quantitative rather than qualitative

Incidence

This is a rare anomaly indeed Ferencz and her co workers[114] were able to collect only thirty four cases from the literature to which they added nine of their own We have seen three patients with congenital mitral stenosis within the past ten years

Anatomy

Anatomically the lesion is seldom an isolated one associated endocardial fibroelastosis is usually found in those patients with severe stenosis who survive for more than a few days Simultaneous involvement of the aortic valve and narrowing of the aorta are relatively common Septal defects except for an occasional patent foramen ovale are rare The consensus of almost all cardiologists is that whenever atrial septal defect with mitral stenosis is present (that is Lutembacher s syndrome) rheumatic etiology of the valvular lesion cannot be excluded with certainty The involvement of the mitral valve consists primarily of a deformity of the chordae tendineae with thickening and stiffening of the valve leaflets themselves

Physiology

The hemodynamic consequences of congenital mitral stenosis are essentially the same as those outlined in the section on rheumatic mitral stenosis (p 151) If a patent ductus arteriosus is associated with severe congenital mitral stenosis the pulmonary vascular obstruction secondary to the severe mitral block may result in a right to left shunt through the ductus

Clinical Picture

The clinical picture of congenital mitral stenosis varies considerably depending on the severity of the lesion Most infants with severe disease

have symptoms within the first few months of life occasionally a child may be free from symptoms for several years Shortness of breath, underdevelopment and mild cyanosis are the most typical presenting complaints

On physical examination underdevelopment is the most striking feature severe cyanosis is absent Mild left chest prominence and a xiphoid type of impulse are usually present Evidences of left sided or right sided congestive failure may be present depending on the severity of the disease In our patients mitral mid diastolic and presystolic murmurs were noted at auscultation but these were not present in the cases described by Ferencz and her associates [114] Accentuated first and second heart sounds are common

The most characteristic roentgenologic finding is the presence of a prominent left atrium in the right anterior oblique view with evidences of significant pulmonary plethora Cardiac enlargement is only moderate and involves the right ventricle in addition to the left atrium (Fig 263)

The electrocardiogram shows right ventricular hypertrophy with or without left atrial hypertrophy

Cardiac catheterization data are available for only one of our patients This child was an older girl in whom the congenital nature of the anomaly could not be ascertained definitely a similar case has been reported by Lurie and Shumacker [8] In both cases the hemodynamic findings could not be distinguished from those associated with moderately severe rheumatic mitral stenosis

In the typical case angiocardiography demonstrates a holdover of the dye in the left atrium for several seconds (Fig 264) This type of examination may also be useful in estimating the size of the left ventricular cavity and thus determining the practicality of mitral valvuloplasty

Course and Prognosis

The prognosis for patients with congenital mitral stenosis varies considerably, depending on the severity of the stenosis and the associated defects Infants with the severest stenosis and left ventricular and aortic involvement resemble patients with an underdeveloped left ventricle and aortic atresia in that they usually die within the first few weeks of life with pulmonary edema and congestive failure Others with relatively minor degrees of stenosis may live for several years without severe disability

Differential Diagnosis

The differential diagnosis of congenital mitral stenosis is extremely difficult often impossible

Rheumatic Mitral Stenosis The only way rheumatic mitral stenosis may be separated from congenital mitral stenosis at present is by con

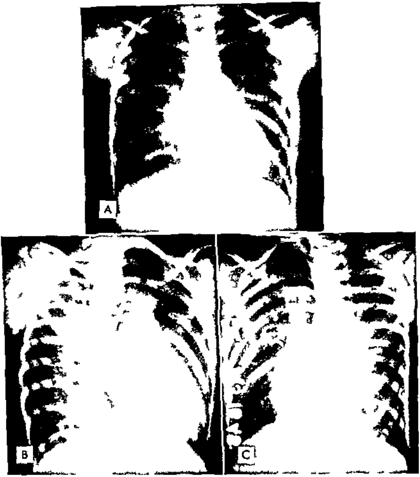


Figure 263 Posteroanterior and oblique radiograms of patient with congenital mitral stenosis Note evidences of left atrial enlargement in the posteroanterior and the right anterior oblique views

sidering the age at which heart disease was discovered However since cases of rheumatic fever have been reported recently in young infants [0] even this landmark may prove to be less reliable than was thought previously

Underdeveloped Left Ventricle with Aortic and Mitral Atresia Patients with congenital mitral stenosis are distinguished from those with underdeveloped left ventricle with aortic and mitral atresia by the fact that in the latter there is severe cyanosis and a large heart and the fact that death occurs within the first few days of life

Endocardial Fibroelastosis Endocardial fibroelastosis without valvular involvement cannot be distinguished from congenital mitral



Figure 264 Angiocardiogram in a patient with congenital mitral stenosis Note the sharp visualization of the left atrium The dye lingered in this chamber for several seconds (Angiogram courtesy of Dr O Hostermann Columbus O)

stenosis unless the characteristic murmur of the latter appears at an early age

Pulmonary Vascular Obstruction *Pulmonary vascular obstruction* should always be considered in the differential diagnosis Primary pul monary vascular disease is best identified by the absence of both left atrial enlargement and characteristic mitral murmur

Constrictive Band across the Left Atrium A constrictive band lying across the left atrium gives rise to high pulmonary venous pressure above the band and relatively normal pressure between the band and the mitral orifice This is an extremely rare anomaly It may be thought of if with a clinical picture of congenital mitral stenosis no left atrial enlargement is present

Treatment

Congenital mitral stenosis may be treated surgically if the condition is uncomplicated and if coexisting endocardial fibroelastosis does not make all attempts at increasing left ventricular output futile Bower and his associates[46] reported two cases in which surgical repair was at tempted one patient survived

At present in my opinion if the patient cannot be substantially improved by medical treatment operation should be attempted *in early* infancy However if the child can survive for a number of years mitral valvuloplasty *should be postponed until later* using the indications out

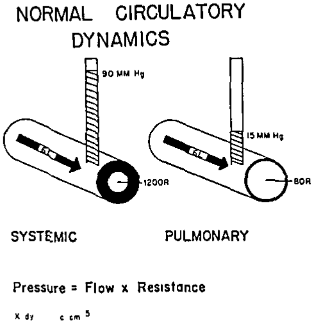


Figure 265 Normal pressure relationship between systemic and pulmonary circuits Note that the pressure in the pulmonary circuit is only a fraction of that in the systemic circuit Corresponding to this the resistance in the pulmonary circuit is lower than the systemic circuit, whereas the flow obviously in the two circuits is identical[90]

lined in the section on rheumatic mitral stenosis (p 155) Results of operation are not known because of the small number of well documented cases

Summary

Congenital mitral stenosis should be suspected in an infant who has evidences of pulmonary vascular obstruction a large left atrium and a mitral diastolic murmur The prognosis is poor in most instances Operation may be attempted

THE PULMONARY VASCULAR OBSTRUCTION SYNDROME

General Principles

The pulmonary arterial systolic pressure of normal persons is about one fourth or one fifth of the systemic arterial systolic pressure (Fig 265) A significant elevation of pulmonary arterial pressure may be the result of either a marked increase in pulmonary blood flow or a significant increase in pulmonary arteriolar resistance with normal or even diminished pulmonary blood flow (Fig 266) In the pulmonary vascular obstruction syndrome the pulmonary arterial systolic pressure is elevated

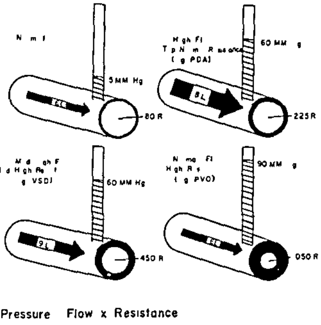


Figure 266 Pulmonary hypertension and its relation to resistance and flow In the upper left hand corner the normal pulmonary vascular bed with normal flow normal resistance and normal pressure is represented In the upper right corner pulmonary hypertension is based on high flow In the lower left corner pulmonary hypertension is based on an increase of flow and resistance whereas in the lower right corner the hypertension is based entirely on increased resistance [9]

to levels identical with or closely approximating the systemic arterial pressure because of a marked increase in pulmonary vascular resistance Thus this syndrome becomes a physiologic entity with good correlations between the calculated resistance and the actual histologic picture of the pulmonary vasculature [90] At our clinic this large increase in pulmonary vascular resistance is designated as the pulmonary vascular obstruction syndrome regardless of whether or not it is associated with any other congenital cardiac anomaly However cases in which the pulmonary vascular disease is clearly secondary to another entity such as mitral stenosis kyphoscoliosis or parenchymatous pulmonary disease are not included in this syndrome Looking at it in still another way we include in the pulmonary vascular obstruction syndrome all patients with "essential pulmonary hypertension[101 364] as well as all patients with congenital heart disease and markedly elevated pulmonary resistance

Incidence

The incidence of this condition cannot be estimated with any accuracy at this time If only the patients with "essential or primary

pulmonary hypertension were considered the incidence would be low—not more than 1 per cent of the patients with congenital heart disease If however patients with congenital heart disease and pulmonary vascular obstruction are included (which we consider reasonable) the incidence may easily become ten times as high This syndrome occurs more often in females than in males

Anatomy

Anatomically speaking the small and medium sized arterioles of the lungs are the site of maximal involvement These vessels show varying degrees of intimal and medial thickening atheromatous plaques and thrombi in various stages of organization The most common cardiac anomaly associated with these vascular changes is a defect of the ventricular septum although patent ductus arteriosus coarctation of the aorta and in older people even atrial septal defect may also occur

Physiology

Physiologically as pointed out earlier the greatly increased pulmonary resistance dominates the picture figures calculated from the pressure gradients across the pulmonary bed and the pulmonary blood flow *show that the pulmonary resistance is equal to or higher than the* systemic resistance This increase in pulmonary arterial resistance causes marked pulmonary and right ventricular hypertension and thus increases appreciably the work of the right ventricle and occasionally even that of the right atrium If the pressure in the right side of the heart is markedly elevated a right to left shunt occurs either through a septal defect or through a forcibly opened foramen ovale

Clinical Picture

Mild degrees of pulmonary vascular obstruction in young children may cause no symptoms at all In older children with more severe disease exertional dyspnea, transient or permanent cyanosis chest pain hemoptysis and syncope are noted Squatting rarely occurs

On physical examination the majority of these patients are fairly well developed Cyanosis of mild degree with slight clubbing is frequently though not invariably present The cardiac impulse is forceful and is localized at the xiphoid process At auscultation the most outstanding finding is the greatly accentuated, even booming rather narrow, second sound at the pulmonary area A grade 2 to 3 soft blowing systolic murmur at the left sternal border is present in the majority but not all of the patients Patients with maximal pulmonary vascular obstruction may have an early insufficiency blow at the left sternal border (Fig 267) No mid diastolic shunt rumble is audible at the apex in these children

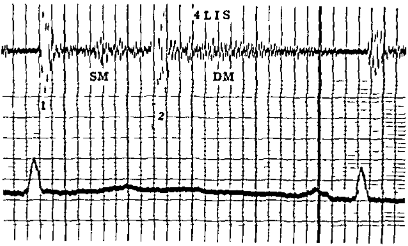


Figure 267 Phonocardiogram in a patient with marked pulmonary vascular obstruction Note the relatively unimpressive systolic murmur and the long loud diastolic murmur at the fourth left interspace

Roentgenologic examination reveals a moderate degree of right ventricular enlargement and a prominent main pulmonary arterial segment (Fig 268) The pulmonary vasculature characteristically shows accentuated hilar markings with diminished peripheral vasculature A hilar dance may be present at fluoroscopy

The electrocardiogram shows varying degrees of right ventricular hypertrophy with moderate P pulmonale (Fig 269)

Cardiac catheterization (Fig 270) reveals the following data (1) There is slight if any evidence of a left to right shunt at the atrial ventricular or pulmonary arterial level (2) There is right ventricular and pulmonary arterial hypertension in the systemic range (3) The "pulmonary capillary" pressure is normal or slightly elevated with a large pulmonary artery pulmonary capillary gradient The brachial arterial tracing is normal (4) Mild or moderate arterial unsaturation is present (5) The pulmonary flow is only slightly if any larger than the systemic flow Pulmonary resistance may be equal to or may even surpass systemic resistance

The risk of cardiac catheterization in this group of patients is higher than average

Occasionally angiocardiography reveals a right to left shunt through a ventricular septal defect It also demonstrates clearly the large pulmonary arteries On the whole however it is not a tremendously informative procedure in this condition and it is also accompanied by appreciable risks

Course and Prognosis

The general outlook for patients with the pulmonary vascular ob

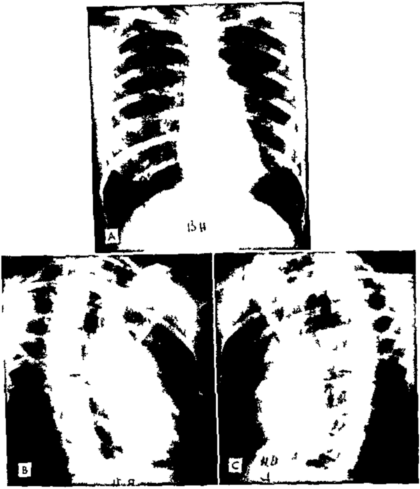


Figure 268 Posteroanterior and oblique radiograms of patient with marked pulmonary vascular obstruction

struction syndrome cannot be described simply because of the varied etiologic factors concerned In cases in which pulmonary vascular obstruction is associated with a defect of the ventricular septum the commonest course of events is that already described in the section on large ventricular defects (p 318) The large left-to-right shunt with high pulmonary arterial pressure causes progressive pulmonary vascular changes and finally obstruction[3 1] Evidences of underdevelopment and congestive failure dominate the picture in infancy This is followed by a period of relative well being with increasing pulmonary resistance which lasts throughout the first and most of the second decade Finally there is a rather rapid downhill course in the late teens and twenties with signs and symptoms of cyanosis and congestive failure

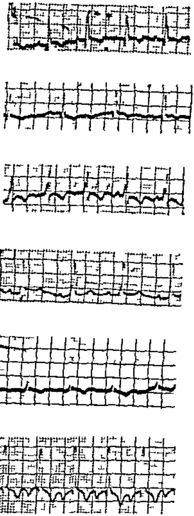


Figure 269 Lead V_1 in six cases of pulmonary vascular obstruction syndrome [90]

In contrast to these patients in whom there is a gradual development of the pulmonary vascular obstruction syndrome from a large left to right shunt are those patients whose pulmonary vascular obstruction exists from early childhood and is truly primary These children may or may not have an associated congenital cardiac anomaly Slight cyanosis exertional dyspnea and underdevelopment exist from infancy No loud murmur particularly no apical diastolic murmur is ever present Cardiac enlargement is always slight Definite right ventricular hypertrophy is shown in the electrocardiogram from the beginning This clinical picture may persist for several years into the first and second decades without significant change Then in the teens or early twenties something happens the beginning of catamenia pregnancy or a severe pulmonary infection may start the patient on a downhill course with increasingly

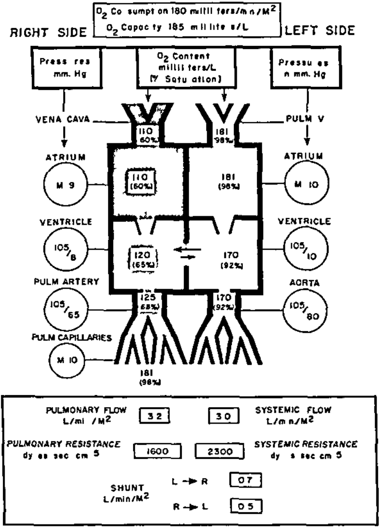


Figure 270 Diagrammatic presentation of findings at cardiac catheterization in patient with pulmonary vascular obstruction syndrome

severe dyspnea on exertion cyanosis syncope and chest pain Palliative measures may prolong life for a few years

A third group of patients will be mentioned only briefly since it consists almost entirely of adults In these pulmonary vascular obstruction starts suddenly without any evidence of cardiac or pulmonary disease in the past In other words these are people who have not previously had a left to-right shunt or evidences of early congenital pulmonary vascular obstruction According to Castleman and his associates [63] many of these patients have primarily thrombo embolic disease

of the pulmonary circuit According to Wood [364] the average survival of these patients after the onset of symptoms is two years

One further point to be mentioned in the prognosis of these patients is the fact that sudden death is relatively common It usually occurs in a syncopal attack perhaps associated with convulsions and probably due to sudden decrease of the cardiac output as a result of increased pulmonary vascular obstruction Also I have seen death occur during cardiac catheterization angiography bone marrow biopsy and even while the circulation time was being determined by injection of Decholin

Differential Diagnosis

The following entities are the principal ones to be considered

Large Left-to-Right Shunt with or without Pulmonary Arterial Hypertension The group as a whole may be identified on the basis of marked cardiac enlargement uniformly plethoric lung fields hyperdynamic cardiac impulse left chest deformity and apical diastolic rumble Individual members of the group (atrial defect ventricular defect or patent ductus arteriosus) can usually be recognized clinically by the characteristic auscultatory phenomena electrocardiograms and specific radiologic contours Cardiac catheterization is frequently necessary to confirm the diagnosis

Valvular Pulmonary Stenosis with Patent Foramen Ovale Valvular pulmonary stenosis with patent foramen ovale may simulate the clinical picture of pulmonary vascular obstruction syndrome in many respects Exertional dyspnea and mild cyanosis may constitute the presenting symptoms in both groups It may be impossible to distinguish between the radiograms or the electrocardiograms in the two conditions However the physical examination revealing the characteristic stenotic murmur at the left sternal border with an absent or decreased pulmonary second sound usually establishes the diagnosis of pulmonary stenosis Obviously the surest diagnostic criterion of pulmonary stenosis is the demonstration of low pulmonary arterial pressure with a gradient across the right ventricular outflow tract

Pulmonary Vascular Obstruction Secondary to Parenchymatous Pulmonary Disease Pulmonary vascular obstruction secondary to parenchymatous pulmonary disease may be diagnosed by appropriate pulmonary function studies

Pulmonary Vascular Obstruction Secondary to Mitral Stenosis It may be extremely hard to differentiate pulmonary vascular obstruction secondary to mitral stenosis from primary pulmonary vascular obstruction In the absence of a good history and the characteristic auscultatory phenomena increased pulmonary capillary pressure and roentgenologic evidence of a large left atrium may be most helpful in the diagnosis With all these diagnostic clues however severe mitral stenosis still may not be recognized by even the most competent people If the exist

ence of mitral stenosis associated with pulmonary vascular obstruction cannot be excluded with certainty left heart catheterization is certainly indicated Fortunately this problem seldom occurs in children, in whom tight mitral stenosis rheumatic or congenital, is exceedingly rare

Pulmonary Vascular Obstruction Secondary to Kyphoscoliosis Pulmonary vascular obstruction secondary to kyphoscoliosis is a rare condition in children If it occurs the spinal deformity is so marked that its identification even without orthopedic assistance is easy

Treatment

Unfortunately no definitive treatment is available for patients with the pulmonary vascular obstruction syndrome A number of palliative measures will be mentioned briefly with the understanding that nothing spectacular can be expected from them at best the progression of the condition may be temporarily halted.

The avoidance of fatigue is important in the management of these children Plenty of sleep at night and at least one or two hours of rest in the afternoon are mandatory Prompt antibiotic treatment for and even continuous chemoprophylaxis against, respiratory disease are recommended. The use of Gantrisin for chemoprophylaxis and one of the broad spectrum antibiotics for treatment is advisable Inhalation of oxygen for thirty minutes two or three times a day has been recommended without much proof of its efficacy The intramuscular administration of Priscoline an antispasmodic, has been recommended by Dresdale and his co-workers[101] to reduce the pulmonary vascular resistance At The Childrens Medical Center we have tried this at cardiac catheterization in a few instances with no appreciable success Long term anticoagulant therapy with Dicumarol to prevent thrombo-embolic phenomena by keeping the prothrombin time at about 30 per cent of normal, has been tried in a few extremely sick patients the results are difficult to evaluate

The presence of an operable congenital cardiac anomaly poses the difficult question Should this defect be repaired? If an atrial defect or a patent ductus arteriosus without appreciable left to-right shunt is present, the answer should be positively "No" My lack of experience about closure of ventricular septal defects forbids me to express a definite *opinion about operation on these lesions in the face of pulmonary vas* cular obstruction I should suspect, however that it might be fraught with just as much danger as if not more than the closing of the other defects mentioned.

Summary

Pulmonary vascular obstruction is characterized by clinical and physiologic evidences of right ventricular and pulmonary arterial hyper tension secondary to severe increased pulmonary arteriolar resistance

Exertional dyspnea cyanosis syncope and chest pain are the dominant symptoms A booming pulmonary closure without a pronounced murmur roentgenologic evidence of moderate right ventricular enlargement and electrocardiographic evidence of severe right ventricular hypertrophy are the most important clinical features The condition may or may not be associated with congenital heart disease Long term prognosis is poor and no adequate treatment is available

PULMONARY STENOSIS

Until the introduction of the cardiac catheter pulmonary stenosis was entirely an anatomic concept According to this definition patients whose right ventricular outflow tract was visibly obstructed had pulmonary stenosis With the advent of cardiac catheterization the old anatomic definition of pulmonary stenosis was supplanted by a physiologic one—patients who have a systolic gradient between the right ventricle and the pulmonary artery (that is patients in whom the right ventricular systolic pressure is appreciably higher than the pulmonary arterial systolic pressure) are now considered to have pulmonary stenosis This concept includes all patients with anatomic stenosis and in addition a large group of patients in whom the usual pathologico anatomic methods fail to show an obstruction

Incidence

Thus defined pulmonary stenosis is present in 20 to 25 per cent of all patients with congenital heart disease In about half of these the ventricular septum is closed in the others a ventricular septal defect with more or less overriding of the aorta is present

Anatomy

Pulmonary stenosis may involve either the semilunar cusps themselves or the subvalvular, infundibular portion of the right ventricle The purely valvular stenoses are usually associated with an intact ventricular septum while as a rule lesions involving the infundibulum are combined with a ventricular septal defect and even overriding of the aorta These rules are not without exception valvular stenosis with a ventricular septal defect and isolated infundibular stenosis do indeed occur

Physiology

The principal physiologic handicap in pulmonic stenosis is the difficulty in maintaining an adequate pulmonary blood flow across a reduced pulmonary orifice In order to accomplish this the right ventricle must work harder and produce much more than the usual amount of pressure If an opening exists between the two sides of the heart and if the right ventricular outflow resistance equals or supersedes the total systemic resistance a right to left shunt with consequent cyanosis results

Because of the difference in symptoms signs and prognosis patients with pulmonary stenosis who have an intact ventricular septum will be discussed separately from those in whom pulmonary stenosis is associated with an open ventricular septum

Pulmonary Stenosis with Intact Ventricular Septum

This anomaly has been variously described as pure pulmonary stenosis isolated pulmonary stenosis" and even pulmonary stenosis with a normal aortic root Since the purity of the lesion is frequently contaminated and its splendid isolation is commonly invaded by an opening in the atrial septum and because the position of the aortic root is difficult if not impossible to define in vivo I prefer the simple anatomic description used in the heading

Incidence

Pulmonary stenosis with intact ventricular septum is a common disease although it was not recognized as such until the past decade In 1947 Taussig[3 1] in her excellent monograph stated that she had not had the opportunity to study a proved case herself and Currens and his associates[89] collected the first large" series consisting of eleven patients in 1945 In contrast to this at The Children s Medical Center in Boston we have studied fifty patients in detail by cardiac catheterization and have seen at least three times that many in whom the diagnosis was made on clinical grounds Wood found twenty four patients with this condition among the first 200 patients with congenital heart disease whom he studied[36] Hence the conservative estimate may be made that patients who have pulmonary stenosis and an intact ventricular septum constitute at least 10 to 15 per cent of all patients with congenital heart disease who live beyond infancy The reason for this apparent change in incidence is hard to explain In my opinion it is due to a clearer definition of the clinical profile revealed by cardiac catheterization probably these patients were formerly classified as having Roger s disease or atrial septal defect

Anatomy

Approximately four of five patients with this anomaly have valvular stenosis[191 311] The cusps are usually fused into a membrane with a hole in the middle which varies in size The excellent angiocardiographic studies of Kjellberg and his associates[191] clearly demonstrate this dome shaped pulmonary valve protruding into the pulmonary artery during systole and collapsing back into the valve ring in diastole If the stenosis is subvalvular—a rather rare occurrence—a small infundibular chamber is usually present beginning several centimeters below the pulmonary valve, the walls of this chamber apparently contract during systole, but fail to open completely in diastole

The right ventricular wall is thickened and the main pulmonary artery is large and thin walled. A clear explanation of the origin of this poststenotic dilatation of the pulmonary artery is contained in a classic paper by Emile Holman.[168] He demonstrated conclusively that the increased lateral pressure resulting from the deceleration of the high velocity flow through a small orifice is probably responsible for this phenomenon.

Embryologically valvular stenosis is thought to be the result of superimposed disease—possibly an inflammatory process—on normally developed valves. To my knowledge there is no definite proof as to the validity of this statement. It becomes a particularly tenuous hypothesis if, as we have had occasion to observe, two or three siblings show the same anomaly.

Physiology

The principal physiologic consequence of pulmonary stenosis with intact ventricular septum is, as mentioned before, a diminution of the pulmonary blood flow. However, there are a few points worth discussing which clearly distinguish this variety of pulmonary stenosis from the hemodynamic viewpoint from other types of block (such as tetralogy of Fallot). The most important consideration is the fact that for a comparable degree of pulmonary stenosis patients with an intact septum may be able to establish a better pulmonary blood flow (and thus better exercise tolerance) than those who have pulmonary stenosis associated with a ventricular septal defect. This is accomplished at the price of a higher right ventricular pressure work which the right ventricle is able to produce by virtue of its independence from the systemic circuit. As a consequence of this markedly increased pressure work, severe right ventricular hypertrophy may be present and eventually, if the stenosis is severe enough, cardiac dilatation with congestive failure may ensue. Cyanosis is a relatively late occurrence in these patients and may be the consequence of one of two mechanisms. It is either due to greatly diminished cardiac output and an increased oxygen extraction at the capillary level (peripheral cyanosis, p. 275) or is the result of a right to left shunt (central cyanosis, p. 275) through an atrial opening (atrial septal defect or patent foramen ovale) made possible by an appreciable increase in right atrial pressure and thus indicating high right ventricular and diastolic pressures.

Clinical Picture

The clinical picture as described in the paragraphs that follow is based principally on data concerning fifty proved cases of pure pulmonary stenosis.[311] A considerable number—at least one fourth—of these patients are completely asymptomatic. Dyspnea and fatigue dominate the picture in the others, whereas cyanosis seems less severe and less fre-

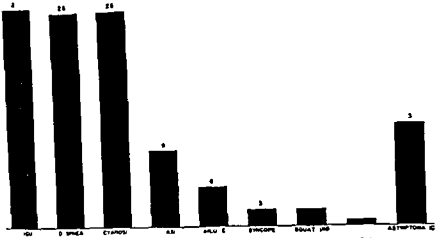


Figure 271 Symptomatology of patients with pulmonary stenosis and intact ventricular septum [311]



Figure 272 Typical appearance of a patient with severe pulmonary stenosis and intact ventricular septum This patient had been operated upon as evidenced by the bandages across the chest Note the chubby round face and good fat deposit over the legs

quent Epistaxis syncope and squatting occur though not commonly (Fig 271) Only rarely are symptoms present in infants most patients do not have complaints until they reach the age of two to five years

On physical examination these youngsters are usually well developed and well nourished the wide pentagon shaped face with high colored cheeks and lips is almost recognizable at the bedside (Fig 272) Definite cyanosis is present in no more than one third of the patients Clubbing if present, is mild On careful inspection the veins in the neck may show a prominent A wave A thrill is commonly present at the suprasternal notch as well as at the upper left sternal border The cardiac impulse is powerful and sustained and is maximal at the xiphoid process Left chest prominence is rare

The auscultatory phenomena are dominated by the stenotic systolic murmur (Fig 273) at the second to fourth left interspace with excellent transmission to the neck and back It is interesting to note that this

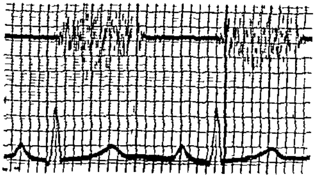


Figure 273 Phonocardiogram of a patient with pulmonary stenosis and intact ventricular septum Note the diamond shaped systolic murmur and the single loud second sound corresponding to aortic closure which is enveloped by the murmur [311]

murmur is usually discovered early often at birth and almost certainly within the first month of life The first heart sound is usually unimpressive although a late systolic click probably due to the jet of blood hitting the thin wall of the pulmonary artery may be noted at the second to third left interspace As a rule the second sound is diminished throughout in the severest stenoses it seems completely obscured by the murmur at the upper left sternal border and is faint elsewhere In cases of mild stenosis the second sound is audible but diminished in intensity at the pulmonary area

The nature of the splitting of the semilunar elements of the second sound in patients with pure pulmonary stenosis may be worth considering in some detail As mentioned earlier in the section on phonocardiography (p 98) the physiologic splitting of the second sound in children is due to the delayed closure of the pulmonary valve If pure pulmonary stenosis of some severity is present the right ventricular systole lasts longer than usual thus the splitting widens On the other hand because of the low pulmonary arterial pressure the pulmonic closure may be faint—even inaudible hence the auscultatory illusion of an unsplit second sound in patients with severe pure pulmonary stenosis (Fig 274) Finally in patients with severe pulmonary stenosis with intact ventricular septum the systolic murmur often extends beyond the aortic closure as far as the closure of the pulmonary valve this may result in the auscultatory impression of an early diastolic murmur at the pulmonary area (Fig 274) A fourth heart sound at the apex is noted in patients with the most severe stenosis and marked right atrial hypertension (Fig 275)

Evidences of congestive failure are rare Infants with the severest disease and children around the age of puberty who have progressively severe stenosis may show cardiac failure In these children presystolic pulsation of the liver may be noted (Fig 276)

Roentgenologically slight or moderate right ventricular enlargement

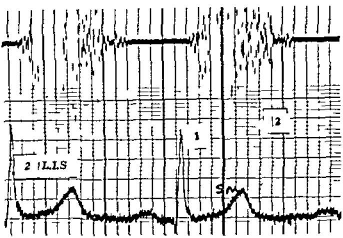


Figure 274 Phonocardiogram of a patient with severe pulmonary stenosis and intact ventricular septum Note the diamond shaped systolic murmur and the wide splitting of the second sound Also note the low intensity of the second component of the second sound

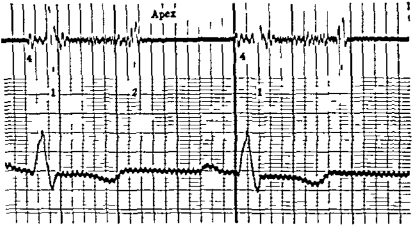


Figure 275 Phonocardiogram at the apex of a patient with pulmonary stenosis and intact ventricular septum Note the loud fourth sound with rather unimpressive systolic murmur at this location

is seen in the great majority of these children Some patients with the *mildest degrees of obstruction have normal sized hearts whereas infants* and children in congestive failure may have unusually big hearts (Fig 277) The enlargement involves principally the right ventricle and the right atrium prominent main and left main pulmonary arteries indicate valvular obstruction Moderate or severe diminution of the pulmonary vasculature is seen in about three fifths of the patients At fluoroscopy the contractions of the heart are powerful except in patients with congestive failure The prominent main pulmonary artery may show pulsa

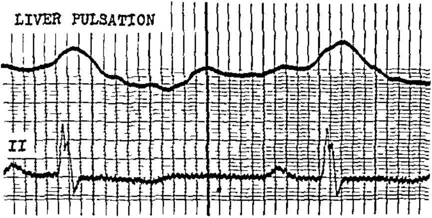


Figure 276 Presystolic pulsation of the liver in patient with severe pulmonary stenosis and intact ventricular septum Liver tracing on top Electrocardiogram tracing as reference tracing on bottom

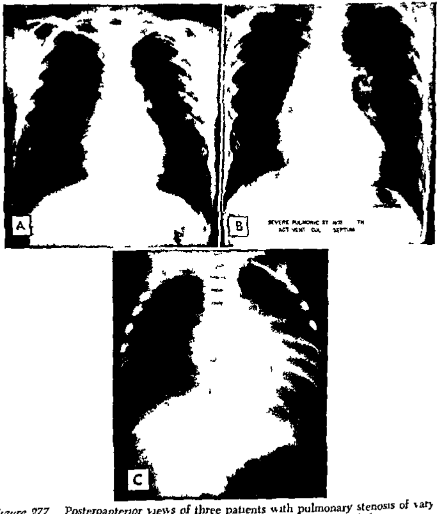


Figure 277 Posteroanterior views of three patients with pulmonary stenosis of varying severity *A* Mild *B* severe *C* very severe in congestive failure

tions but no expansile pulsation of the smaller pulmonary vessels is ever seen

In all patients with moderate or severe stenosis the electrocardiogram shows right ventricular hypertrophy only ten of the fifty patients studied in detail had normal tracings Incomplete right bundle branch block is present in a considerable number of children it dominates the picture in those with mild pulmonary obstruction The tracings with the severest degrees of right ventricular hypertrophy are prone to show significant P pulmonale and changes in the S T segment and the T wave from $V_{1\ 4}$ or even V (Fig 278)

Cardiac catheterization reveals the following data (1) A small left to right shunt at the atrial level may be observed in patients who have an atrial septal defect and mild stenosis

2 A gradient exists between the right ventricular and pulmonary arterial systolic pressures The systolic pressure within the right ventricle is always elevated varying from approximately 50 to 250 mm of mercury The pulmonary arterial mean pressure varies within a much narrower range between 10 and 20 mm of mercury (Fig 279) There is usually a substantial difference between the systemic arterial and the right ventricular systolic pressures There is some evidence to the effect that the pulmonary arterial pressure recorded at catheterization may be lower than it actually is, because of the obstruction of the pulmonary orifice by the catheter The site of the pulmonary obstruction may be clearly demonstrated by slowly withdrawing the cardiac catheter from the pulmonary artery to the right ventricle When this is done the tracing may demon trate either a *sudden* jump in systolic pressure at the valve site (Fig 280 *A*) within one cardiac cycle or a gradual increase in systolic pressure indicating the presence of an infundibular chamber Within this chamber the diastolic pressure is identical with that found in the body of the right ventricle (Fig 280 *B*) whereas the systolic pressure is about midway between that of the pulmonary artery and the body of the right ventricle A diagnosis of valvular obstruction made at cardiac catheterization can almost invariably be confirmed at operation The vast majority of the patients with valvular stenosis have poststenotic dilatation of the pulmonary artery which is visible on the roentgenogram The right atrial pressure tracing reveals giant A waves in patients with the severest stenosis

3 The pulmonary capillary and brachial arterial pressures are usually within normal limits

4 Definite arterial unsaturation at rest is present in about 10 per cent of the children always the patients with the highest degrees of obstruction (Table 32) In perhaps another 10 per cent the arterial saturation is between 90 and 94 per cent at rest with an appreciable drop on exercise

5 The cardiac output at rest is at the lower limits of normal in

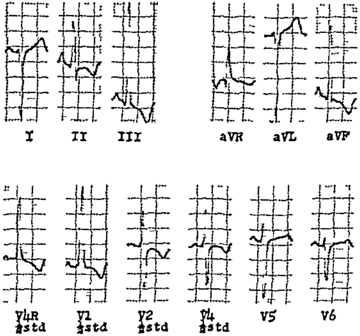


Fig 278 A.

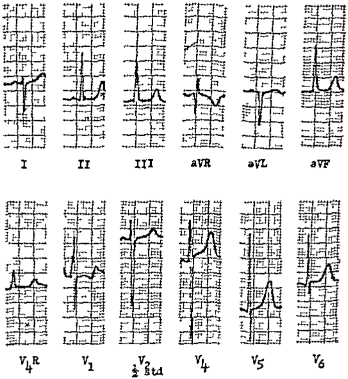


Fig 278 B

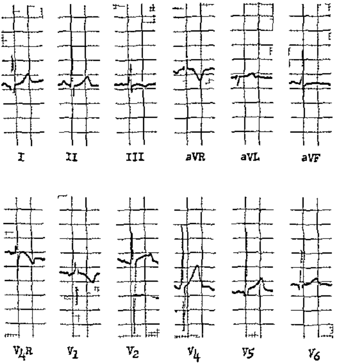


Fig 278 C

Figure 278 Electrocardiograms of patients with pulmonary stenosis *A* Very severe *B* moderately severe *C* mild

practically all patients The expected increase in cardiac output on *exercise is inadequate in the severe cases and is accomplished principally* by widening the arteriovenous difference The pulmonary valve resistance is moderately or markedly elevated

Usually it is not necessary to perform cardiac catheterization to establish the diagnosis in these patients clinical methods are thoroughly satisfactory in this respect At present cardiac catheterization is recommended in instances in which the clinical diagnosis is not clear cut or preoperative accurate appraisal of the severity of the stenosis is desirable

If an attempt is made to correlate the cardiac catheterization data with the clinical picture in this group of patients (*Fig 281*) interesting and useful clues are revealed Patients with a right ventricular pressure of more than 100 mm of mercury cannot be distinguished easily from those with lower pressures by the character of the murmur the presence of a thrill the intensity of the second sound at the pulmonary area or the radiologic evidences of right ventricular enlargement They may be separated with somewhat more accuracy by taking into consideration the presence of symptoms and the degree of pulmonary ischemia revealed by roentgenogram Only by analyzing the electrocardiogram

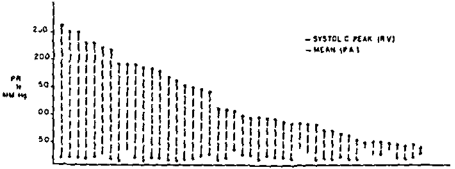
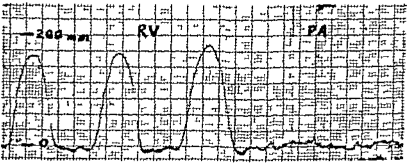
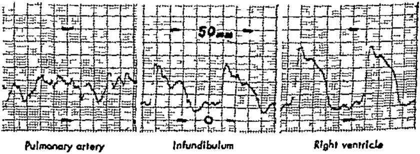


Figure 279 Pressure gradients in forty five patients with pulmonary stenosis with intact ventricular septum Systolic peak pressure top mean pulmonary arterial pressure bottom line Note the relatively narrow fluctuation of the pulmonary arterial mean pressure and the wide variation of the right ventricular peak systolic pressure [311]

A



Isolated pulmonary stenosis valvular type Pressure tracing machine speed 50 mm /sec



B

Isolated pulmonary stenosis infundibular type Pressure tracing machine speed 50 mm /sec

Figure 280 Pressure tracings during a pull back of the catheter from pulmonary artery into right ventricle *A* The sudden jump occurring within one cardiac cycle characteristic of valvular pulmonary stenosis *B* Gradual change in systolic pressure in the presence of an infundibular chamber [311]

Table 32 Correlation of Right Ventricular Systolic Peak Pressures with Arterial Unsaturation and with Presence of Giant "A" Waves

Right Ventricular Pressure (mm Hg)	*200–250*	*150–199*	*100–149*	*50–99*	*Under 50*
Distribution of patients	7	8	5	13	12
No with saturation under 90%	3	3	—	—	—
No with saturation 90–94%	—	2	1	2	—
No with giant A wave	7	5	2	—	—

* Forty five catheterized patients with pulmonary stenosis and intact ventricular septum (42 valvular stenosis 3 infundibular stenosis) [24]

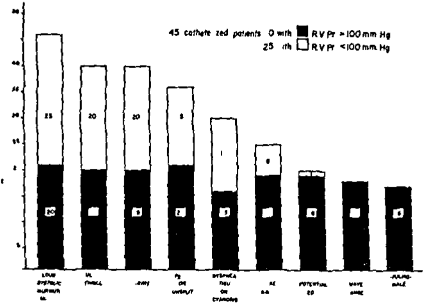


Figure 281 Correlation of right ventricular systolic peak pressure with the clinical picture in forty five patients with pulmonary stenosis and intact ventricular septum For explanation see text [311]

carefully however *can one determine with relative certainty* on clinical grounds whether the patient's right ventricular pressure is higher or lower than 100 mm of mercury We have not found any patient with a right ventricular pressure of more than 100 mm of mercury who has not demonstrated at least one of the following three important electrocardiographic criteria (*1*) R_{V1} of more than 20 mm (*2*) P pulmonale and (3) changes in the S T segment and the T wave through V_4 Furthermore once any of these features is noted the chances that the pressure in the right ventricle is high are good As can be seen in Figure 281 there was only one patient with an R wave in V_1 of more than 20 mm who did not have a right ventricular pressure of more than 100 mm of mercury *Important as these correlations* between right ventricular



Figure 282 Angiocardiogram outlining main pulmonary artery and left main pulmonary artery with tremendous poststenotic dilatation in a patient with valvular pulmonary stenosis

pressure and the electrocardiogram are it must be remembered that we can attest for their validity only in infants and children and only in cases of isolated pulmonary stenosis Any conclusions drawn from these figures in relation to other conditions and other age groups would have to be made with the greatest caution

Angiocardiography especially the selective technique practiced by Swedish workers[191] has produced some of the most striking pictures (Fig 282) seen in the study of congenital heart disease As a scientific tool designed to help physicians understand the dynamics of this lesion and the structural changes underlying it angiocardiography has fulfilled its immensely important task For practical purposes however in the diagnosis and treatment of congenital pulmonary stenosis it is of relatively little use today The presence or absence of a ventricular defect may be determined better at cardiac catheterization The cardiac catheter is also more useful than angiocardiography at least in our laboratory in determining the nature of the stenosis Therefore in my opinion although angiocardiography is a magnificent teaching and research instrument in the study of pulmonic stenosis its practical use is limited at present and it is overshadowed by cardiac catheterization

Course and Prognosis

The prognosis in cases of pulmonary stenosis with intact ventricular

septum is difficult to predict accurately at present because of the relatively short observation period available as mentioned earlier large numbers of these patients have not been observed clinically until the past ten years However on the basis of our own observations during these ten years it seems that a number of fairly distinct categories may be outlined

The first group of patients those with the severest disease exhibit evidences of congestive failure within the first year of life Anatomically these patients have pinpoint stenosis of the pulmonary valve but they probably constitute less than 10 per cent of the total number of patients with this disease They are usually well developed babies who have a tinge of cyanosis a large heart and electrocardiographic evidences of severe pulmonary stenosis Unless they are operated on these infants probably die within the first year or two of life

The mild symptoms in the second group of children usually start within the first years of life If the electrocardiographic criteria may be relied on these youngsters have progressively higher right ventricular pressures through the years with increasing fatigue and dyspnea and the late appearance of cyanosis We know of at least one such child who died suddenly at the age of ten years This is a rare occurrence however the usual evolution is one of increasing symptoms evidences of right sided failure and death in the late teens or twenties

The patients in the third and final group do not have any symptoms throughout childhood the right ventricular pressure when measured is less than 100 mm of mercury and the electrocardiogram fails to reveal evidences of progressively increasing right ventricular pressure through the years It is this group of children—the asymptomatic low pressure group—about whom information is woefully lacking particularly in regard to their fate in adulthood At present we do not even know whether the stenosis becomes relatively more or less severe as the years go by Only careful observation through many decades will furnish a definite answer to this question

Among the complications of pulmonary stenosis the rare occurrence of subacute bacterial endocarditis and the rather unlikely appearance of rheumatic fever may be mentioned We have encountered one patient whose sudden catastrophic increase in symptoms was explained at post mortem examination on the basis of an infected vegetation obstructing an already small pulmonary orifice

Differential Diagnosis

Tetralogy of Fallot The most common problem in the differential diagnosis of pulmonary stenosis with intact ventricular septum is to distinguish it from tetralogy of Fallot

The symptomatology of the two conditions shows slight but definite differences In tetralogy of Fallot usually though not invariably cya

Table 33

DIFFERENTIAL DIAGNOSIS OF P.S. $\bar{c}$ INTACT VENTRICULAR SEPTUM & TETRALOGY OF FALLOT

	PULMONARY STENOSIS	TETRALOGY OF FALLOT
SYMPTOMS		
CYANOSIS	±	+
DYSPNEA	+	+
SPELLS	0	+
SQUATTING	±	+
C F	+	0
P.E.		
GIANT A WAVE	−	0
SECOND SOUND AT 2 LIS SPLIT	+	0
SECOND SOUND AT 2 LIS INTENSITY	↓	↓
AT 4 LIS SPLIT	+	0
AT 4 LIS INTENSITY	↓	↑
SYSTOLIC M		
AT 2 LIS	+	−
AT 4 LIS	0	+
EKG		
RVH	++	+
P PULMONALE	+	±
X RAY		
GENERAL CE	±	0
RVE	++	+
PROMINENT PA	+	0
PV	N OR ↓	↓
RAA	0	25%
CARDIAC CATH.		
RV PRESSURE > SYSTEMIC	±	0
GIANT "A" WAVE	−	0
VALV. STENOSIS	+	±

See Table 26 (p 303) for explanation of symbols

nosis and exertional dyspnea are present in almost equal degree at all times while in pure pulmonary stenosis fatigue and dyspnea are much more prominent than cyanosis Squatting and anoxic spells are more common in tetralogy of Fallot than in pulmonary stenosis with intact ventricular septum Congestive failure—a real danger in patients with pure pulmonary stenosis—never occurs in children with tetralogy of Fallot

On physical examination the finding of a giant A wave in the jugular venous pulse indicates pure pulmonary stenosis If the stenotic murmur is heard best high at the left sternal border and especially if it completely obscures the aortic closure the lesion is probably valvular pulmonary stenosis with an intact septum, the presence of a loud narrow

second sound observed at the lower left sternal border strongly suggests tetralogy of Fallot It should also be mentioned that though there may be no murmurs at all in severe cases of tetralogy of Fallot we at The Children s Medical Center still have to see a case of pure pulmonic stenosis in which a stenotic murmur has not been observed although such has been noted by others [33]

The electrocardiogram as a rule shows more severe degrees of right ventricular hypertrophy with strain and more P pulmonale in patients with severe isolated pulmonary stenosis than in those with tetralogy of Fallot

The roentgenologic picture may be considerably different in the two *conditions as well In almost 25 per cent of the patients with tetralogy* of Fallot there is a right aortic arch whereas we have never seen this occur in a patient who has pulmonary stenosis with an intact ventricular septum only one such case has been mentioned in the literature [71] Other important radiologic differences between the two conditions are the frequent presence of a poststenotic dilatation and the occasional occurrence of marked cardiac dilatation in severe pulmonary stenosis whereas in tetralogy of Fallot the cardiac enlargement is always slight to moderate and the main pulmonary artery segment usually concave

The Left to Right Shunt Group Atrial septal defect, ventricular septal defect and patent ductus arteriosus—the left to right shunt group—are commonly mentioned in the differential diagnosis of "isolated" pulmonic stenosis In my opinion the question need seldom if ever arise The tumultuous cardiac impulse the left chest deformity the apical diastolic rumble and the pulmonary vascular engorgement with expansile pulsations all identify the left to right shunt group clearly Individual lesions within the group can usually be identified on the basis of physical electrocardiographic and roentgenologic examinations

The Pulmonary Vascular Obstruction Syndrome The pulmonary vascular obstruction syndrome and pulmonary stenosis with intact ventricular septum are discussed on page 434

Ebstein s Anomaly Ebstein s anomaly should be considered in the differential diagnosis of only the severest degree of pulmonary stenosis with congestive failure and marked cardiomegaly The similarities are largely radiologic both conditions being characterized by a large poorly pulsating heart and diminished pulmonary vasculature The differentiation rests on the following points (*1*) The physical examination in Ebstein s disease fails to show the characteristic stenotic systolic murmur but shows a triple or quadruple apical rhythm with low pitched systolic and diastolic murmurs (2) The electrocardiogram in Ebstein s disease never shows marked right ventricular hypertrophy but rather the curious combination of a great deal of conduction disturbance with marked P pulmonale

Aortic Stenosis See page 406

TREATMENT

The treatment of pulmonary stenosis if any is surgical Two main techniques are recommended today one is Brock's approach [1] to the pulmonary valve through the right ventricular musculature and the other is Swan's technique[40] of splitting the valve from the pulmonary arterial side during complete interruption of the circulation under hypothermia The simplicity of the procedure and the good results obtained in the majority of the instances have persuaded the members of our surgical service to use the former rather than the latter method so far for the relief of pulmonary obstruction [311] A shunt procedure is contraindicated in patients with pure pulmonary stenosis not only does it afford no relief but also it may lead to congestive failure and death (see p 474)

Fortunately most of the patients with severe pulmonary stenosis have a valvular type of obstruction which is relatively easy to correct surgically The only patients we have lost during this operation were those in congestive failure the mortality for the entire group was 10 per cent and was zero for those without congestive failure Symptomatically the postoperative results were gratifying in all instances The right ventricular pressures and the maximal right ventricular potentials in the electrocardiograms preoperatively and postoperatively in eight patients are compared in Figure 283 This figure shows the effectiveness of the operation and the reliability of the electrocardiogram in demonstrating the changes in the right ventricular pressure Figure 284 shows the change in the radiologic picture before and after the operation

Isolated infundibular pulmonary stenosis seldom occurs In my opinion patients with this condition probably should not be operated on until safe methods for open heart surgery and relief of the obstruction under direct vision become available

Indications for pulmonary valvotomy can be summarized as follows (1) Young infants with the severest stenosis and congestive failure should be subjected to operation as soon as the diagnosis is established [13] Cardiac catheterization should be performed only if the cardiologist feels that the baby can tolerate the procedure and the delay of the operation Actually this is an emergency procedure Anticongestive measures may be undertaken but usually they are not successful and may waste precious time

2 In patients with clinical indications of right ventricular pressure of 100 mm of mercury or more cardiac catheterization certainly and probably eventually operation should be performed The optimal timing of operation in this group depends on a number of factors If the patient is symptomatic the decision is easy If the child is asymptomatic the presence of significant or progressive cardiac enlargement progressive

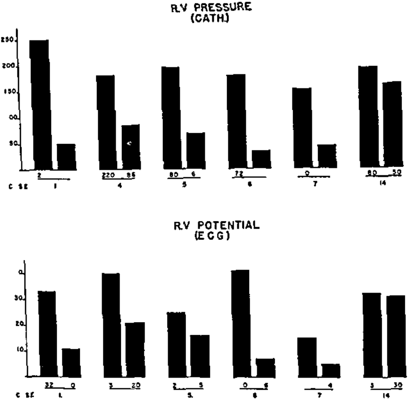


Figure 283 Preoperative and postoperative right ventricular peak systolic pressures and preoperative and postoperative electrocardiograms in six patients with pulmonary stenosis and intact ventricular septum Note the good parallelisms between pressure drops and drop in right ventricular voltages in the electrocardiogram In Case 14 there has been a negligible drop both in electrocardiographic voltages and in right ventricular pressures In all the others a significant drop in both occurred[311]

electrocardiographic changes or electrocardiographic evidences of severe right ventricular strain may be considered valid indications for operation

If operation on a patient of this group is temporarily postponed a follow up examination every three to six months with careful repeated evaluation of the clinical electrocardiographic and roentgenologic data is essential

3 Patients with clinical indications of "pure" pulmonary stenosis and a right ventricular pressure of less than 100 mm of mercury should not be operated on at this time but rather should be examined every six to twelve months to evaluate the progression of the condition Cardiac catheterization is optional in this group of patients As indicated previously these children may have a mild form of the disease which may never need correction The validity of this assumption can be tested only by careful follow up examinations for many years

Among the postoperative complications the development of peri

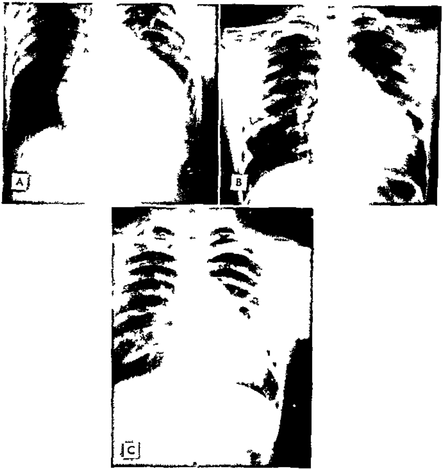


Figure 284 Changes in the heart size of patient with severe pulmonary stenosis and intact ventricular septum *A* The preoperative film *B* a year and a half after operation *C* three years after operation

cardial effusion congestive failure and arrhythmias should be mentioned Medical treatment or surgical drainage of the effusion invariably clears up these complications However careful postoperative supervision is essential for the discovery of these conditions No particular preoperative care is necessary but because of the tendency for arrhythmias to develop during valvotomy administration of digitalis should be stopped at least forty eight hours before the operation

SUMMARY

Pulmonary stenosis with an intact ventricular septum is characterized by a stenotic systolic murmur at the second left interspace diminution of pulmonary closure and evidences of right ventricular hypertrophy and enlargement Patients with the milder form of the disease may be completely asymptomatic others show dyspnea on exertion fatigue and

slight cyanosis Children with valvular stenosis and severe disease should be operated on The electrocardiogram is the best clinical tool for ascertaining the severity of the lesion

Pulmonary Stenosis with Ventricular Septal Defect and Right to Left Shunt (Tetralogy of Fallot)

Pulmonary stenosis or atresia associated with a ventricular septal defect and dextroposed aorta forming the anatomic basis of cyanotic congenital heart disease was described about 180 years ago [01] Almost 100 years later Fallot[11] defined this syndrome as consisting anatomically of four features—pulmonary stenosis or atresia dextroposition of the aorta ventricular septal defect and right ventricular hypertrophy With the introduction of cardiac catheterization the physiology of this anatomicochemical syndrome was revealed The physiologic definition of tetralogy of Fallot is the combination of a ventricular septal defect with pulmonic stenosis in which the shunt is predominantly or exclusively right to left Although the static term "tetralogy of Fallot is being and probably will be used by everyone for many decades my own objection to the term based principally on an aversion to eponyms finds a considerable amount of support *in the many case reports of formes frustes* of this disease barely explainable on strict anatomic grounds I see no logical reason at this time for trying to separate patients within this general category according to the degree of pulmonary stenosis or dextroposition of the aorta All patients who have pulmonary stenosis and ventricular septal defect with a right to left shunt will be discussed in this section

Incidence

The tetralogy of Fallot is a common disease it accounts for at least 15 per cent of all children more than two years of age who have congenital heart disease It is the most common single lesion in patients with cyanotic disease who live beyond infancy If a person not well versed in the mysteries of pediatric cardiology simply made a diagnosis of tetralogy of Fallot in every cyanotic child more than two years old he would probably be right at least 75 per cent of the time

Anatomy

Embryologically the combination of ventricular defect and pulmonary stenosis may be explained by a misplacement of the spiral ridge dividing the truncus arteriosus resulting in unequal division of the common trunk and failure to close the ventricular septum from above This usually occurs in the fifth to seventh week of intrauterine life

According to our experience and that of others [18, 63] the stenosis is infundibular in the vast majority of instances Probably pure valvular stenosis is not associated with tetralogy of Fallot in more than 10

per cent of the cases. However, opinions contrary to this have been expressed by Brock and Bailey,[1] who maintain that the stenosis is valvular in a considerable portion of the patients. The angiographic studies of the Swedish workers[191] show the infundibular chamber to be a long, narrow channel which contracts completely during ventricular systole but fails to relax during diastole.

A defect of varying size in the membranous subaortic portion of the ventricular septum is present in all instances. It is usually crescent shaped, with a concavity upwards. The severity of the so-called overriding of the aorta may be difficult to determine even at postmortem examination. Most of the time a clinical determination of the overriding is impossible. As a matter of fact, because of the nature and position of the high ventricular defect, the aorta communicates with both ventricles in practically every case, according to Selzer.[317]

The degree of right ventricular hypertrophy depends principally on the severity of the pulmonary stenosis; usually the thickness of the right ventricle equals that of the left. The foramen ovale is patent in considerably more than the expected 20 to 25 per cent of these patients, and a right aortic arch is present in 25 per cent. Significant enlargement of the bronchial arteries occurs in patients with extreme tetralogy of Fallot.

Physiology

In every patient with tetralogy of Fallot there exists a systolic pressure gradient between the right ventricle and the pulmonary artery. This results in a pulmonary resistance that approximates or even supersedes the systemic resistance, thus causing a right to left shunt through a ventricular defect and essentially identical systolic pressure levels in the right ventricle and the systemic artery. The size of the right to left shunt may vary, depending on the ratio between the resistances in the two circuits, the size of the ventricular septal defect, and on certain characteristics of the right ventricular contraction.

The handicaps resulting from this anomaly are twofold. First, because of the right to left shunt, central cyanosis of varying degree is present in the vast majority of patients. Second, because of the pulmonary obstruction, which cannot be met with a right ventricular pressure appreciably higher than the systemic arterial pressure (due to the presence of a ventricular defect), the pulmonary blood flow is limited and exertional dyspnea is marked. The presence of the ventricular septal defect thus places a ceiling (that of the systemic arterial pressure) on the pressure exerted by the right ventricle. This fact, although responsible for the marked limitation of exercise, also has the beneficial effect of protecting the right ventricle from doing too much pressure work. This explains the observation that although these patients may be extremely limited in their activities, cardiac failure is almost never seen during childhood.

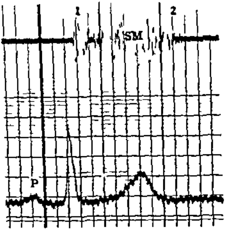


Figure 285 Phonocardiogram of patient with tetralogy of Fallot syndrome Note the diamond shaped systolic murmur ending abruptly with the narrow second sound

Clinical Picture

Few children with tetralogy of Fallot are asymptomatic most of them have the habit of squatting they show evidences of cyanosis and dyspnea on exertion and they are likely to have attacks of deep cyanosis unconsciousness and convulsions

The cyanosis is of the central type and is usually accompanied by clubbing Exercise tolerance varies some patients may be able to walk less than one block, while others may be able to walk a mile or more When these children reach their limit they usually squat for a few minutes and then get up and resume their previous activities The nature of the anoxic spells has already been described on page 277

On physical examination these patients appear moderately under developed they rank between the tenth and the twenty fifth percentile on standard development charts Cyanosis and clubbing of various degrees are evident No giant A waves are visible in the jugular venous pulse Left chest prominence is absent or minimal The cardiac impulse is maximal at the xiphoid process but there is some apical impulse as well A thrill at the lower or mid left sternal border is noted in more than half of the patients

At auscultation the first heart sound is usually narrow and loud at the apex and the lower left sternal border The second sound is almost invariably diminished at the pulmonary area but is narrow (unsplit) and accentuated at the third or fourth interspace parasternally Third sounds or atrial sounds either are not heard or are insignificant It is important to note that the second sound is never completely obliterated by the murmur

The systolic murmur is usually stenotic in character grade 3 to 4 in intensity and is best heard at the mid or lower left sternal border In

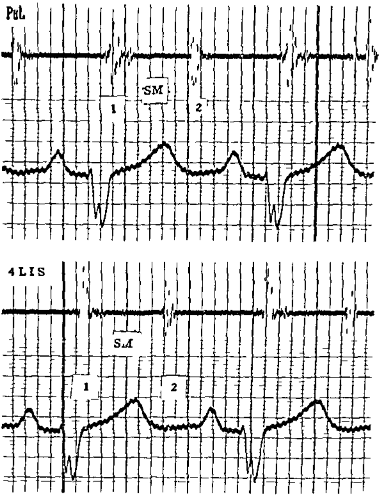


Figure 286 Phonocardiogram in a patient with severe tetralogy of Fallot Note the relatively loud single second sound at the pulmonary area and the fourth left interspace and the slight systolic murmur

contrast to pulmonary stenosis with an intact ventricular septum the systolic murmur never lasts beyond the aortic closure (Fig 285) hence no illusion of an early diastolic blow is created In the most severe cases of tetralogy of Fallot there may be only a slight murmur or none at all (Fig 286) These are the only ones in which practically no blood passes through the pulmonary obstruction It is interesting to note the variability of the systolic murmur in these patients although a child may have a grade 3 systolic stenotic murmur when feeling relatively well at the height of an anoxic spell this same youngster may not have a murmur at all With the improvement of the child s condition the murmur usually returns

These changes in auscultatory findings can be explained on the basis of variable volumes of blood at variable velocities traversing

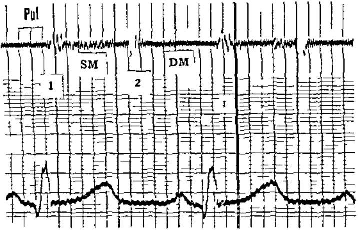


Figure 287 Phonocardiogram in patient with severe tetralogy of Fallot Note the presence of a continuous murmur at the pulmonary area Note the high frequency low intensity vibration and the absence of the crescendo systolic element

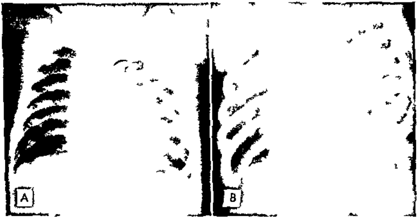


Figure 288 Posteroanterior and left anterior oblique radiograms in patient with tetralogy of Fallot Note the relatively small heart the absent main pulmonary artery segment the relatively wide supracardiac shadow with a dilated superior vena cava the right ventricular contour and pulmonary ischemia

the stenotic right ventricular outflow tract at various times A continuous murmur is often heard in patients in whom there is no stenotic systolic murmur (Fig 287) This is usually heard best at the second right and second left interspaces as well as over the back and can be explained on the basis of the wide network of bronchial collaterals In rare instances the continuous murmur may be caused by a patent ductus arteriosus

The roentgenologic picture (Fig 288) is characterized by a relatively small heart of right ventricular contour and a poorly vascularized

lung field (*coeur en sabot*) If there is any cardiac enlargement shown in the roentgenogram, it involves almost exclusively the right ventricle and perhaps the right atrium The main pulmonary arterial segment is small even concave the aorta on the other hand is large The aortic arch is commonly on the right side The peripheral pulmonary vasculature may be normal or diminished and a pattern of collateral circulation may be noted in patients with the severest pulmonary stenosis

The electrocardiogram is always abnormal A moderate or severe degree of right ventricular hypertrophy is noted with R waves of variable height in the right chest leads No good correlation exists between the QRS voltages and the right ventricular pressure in these patients Evidence of additional left ventricular hypertrophy may sometimes be obtained from leads aV_F and V_{5-6} (Fig. 289)

Cardiac catheterization (Fig 290) reveals the following (1) Only in those patients with the mildest stenosis is a slight left to right shunt at the ventricular level sometimes demonstrated at rest on exercise or crying this shunt is usually abolished and the flow of blood through the ventricular defect is exclusively right to left (2) There is always right ventricular systolic hypertension in the systemic range the presence of a pressure gradient between the right ventricle and the pulmonary artery cannot always be proved at catheterization since the catheter may not be able to enter the pulmonary artery If the pulmonary artery is entered however the presence of infundibular stenosis can be demonstrated on withdrawal (Fig 291) The pulmonary arterial pressure is lower than normal and often shows marked respiratory variations and damping (3) "Pulmonary capillary" and brachial arterial tracings are usually within normal limits (4) The arterial oxygen saturation varies between 30 and 80 per cent because of the right to left shunt through the ventricular septal defect In the presence of a patent foramen ovale an additional right to left shunt may occur at the atrial level (5) A right to left shunt on exercise may be calculated in all instances in the severest cases it may be present even at rest The total pulmonary resistance is equal to or greater than systemic resistance (6) In approximately a third of the patients the left ventricle or the aorta may be entered from the right ventricle

Cardiac catheterization usually is not necessary for the diagnosis of the tetralogy of Fallot In most instances the clinical profile is clear enough to allow accurate diagnosis prognosis and an outline of therapeutic procedures However right heart catheterization is indicated if the clinical diagnosis is not clear or if some specific physiologic information is desired

Angiocardiography is useful in those patients with the tetralogy of Fallot in whom the size of the pulmonary artery is not clearly demonstrable by ordinary radiography but it certainly is not a necessary procedure in the vast majority of cases Selective angiocardiography as

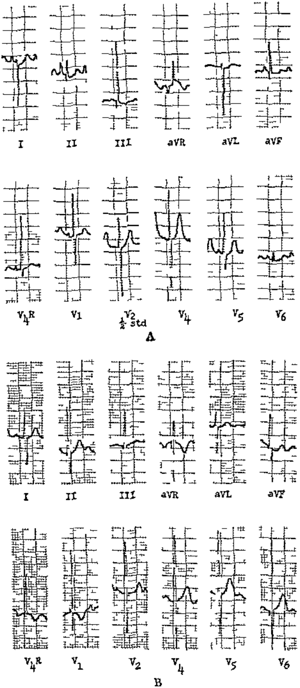


Figure 269 Two electrocardiograms of patients with tetralogy of Fallot syndrome *A* Pure right ventricular hypertrophy *B* Combined ventricular hypertrophy Note the P pulmonale in *A*

lung field (*cocur en sabot*) If there is any cardiac enlargement shown in the roentgenogram it involves almost exclusively the right ventricle and perhaps the right atrium The main pulmonary arterial segment is small even concave the aorta on the other hand is large The aortic arch is commonly on the right side The peripheral pulmonary vascula ture may be normal or diminished and a pattern of collateral circulation may be noted in patients with the severest pulmonary stenosis

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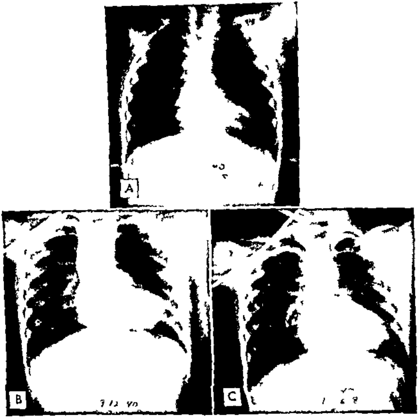


Figure 292 Venous angiograms in patient with the tetralogy of Fallot syndrome A Plain film without injection of the dye B Dye entering the right atrium and right ventricle through the superior vena cava C Visualization of the aorta from the right ventricle and appearance of collaterals from the aorta

discussed in the section on pure pulmonary stenosis is an excellent tool for research for teaching and for the understanding of this malformation This type of examination as well as conventional venous angiography may give practical information preoperatively in regard to the nature of the stenotic area the presence and degree of overriding of the aorta and the presence and size of the pulmonary arteries (Fig 292)

Course and Prognosis

Approximately one third of the patients with the tetralogy of Fallot show cyanosis at birth In another third blueness is first exhibited between the ages of one month and one year The remainder of the patients do not have noticeable arterial unsaturation until they are well past the first year of life A grouping of these patients according to the time of initial appearance of cyanosis is possible and perhaps even useful if the

reader realizes that most patients fall somewhere in between the three main groups

Group 1 is made up of patients who are cyanotic at birth or shortly thereafter They usually have the severest form of the disease and they may be the ones in whom because of the physiologic or anatomic obliteration of the pulmonary orifice no stenotic systolic murmur is audible The absence of any demonstrable flow through the pulmonary orifice and the presence of collaterals with a continuous murmur suggested the name "pseudotruncus" which is applied to these patients At four or five months of age these infants start to have anoxic spells (see p 277) of increasing severity and frequency Unless they are operated on they rarely survive the first year of life Death usually occurs in a major anoxic spell

If they do survive the first twelve to eighteen months of life they may live for several years but are deeply cyanotic and polycythemic with marked limitations of exercise and squatting habits With all this they may be mentally alert able to do school work at home and to occupy themselves in a sedentary fashion They seldom survive the second decade but their brief life span is often an inspiring example of the indomitable nature of the human spirit

Group 2 is made up of patients whose cyanosis becomes obvious only after the first month of life but before the end of the first year This group is regarded as most typical of the tetralogy of Fallot syndrome Anoxic spells also may appear in these patients at or about six months of age but they are neither as numerous nor as severe as those noted in the first group of infants Sometimes they take the form of irritable spells or prolonged crying episodes early in the morning A loud systolic murmur is usually audible at the left sternal border the murmur of collaterals is absent When these children are able to walk (at one and a half to two and a half years of age) dyspnea on exertion and squatting become obvious Symptoms are rather mild in favorable weather but extremes in temperature are poorly tolerated These children may run around and play satisfactorily in the house but when they go out to the street or play area they squat or ask to be picked up after they have walked a few hundred yards A peculiar irritating hacking cough appears when they tire

When school age is reached the difficulties increase Squatting is a socially unacceptable position and dyspnea on exertion makes it impossible for them to play the games all children enjoy The walk to school and the climbing of stairs become insurmountable obstacles Without the help of an operation life becomes less and less tolerable as the years go by Perhaps the disease is harder for these youngsters to tolerate because since they are not as sick as those in the first group they may try to look and act like other children and of course they fail The patients in the first group are so completely different from the average

youngster that they may be able to create a world of their own a task sometimes simpler than attempting to become a member of a group that does not want you Without operation the patients in the second group usually live through the second or even the third decade Death occurs from inadequate cardiac output inadequate oxygen uptake or a cerebrovascular accident Congestive failure may be seen rarely in adults

The third group of patients with tetralogy of Fallot do not show evidences of cyanosis until they are one year old or older These are the children who sometimes are referred to as "acyanotic tetralogies"[365] During infancy and early childhood they may show a characteristic picture of ventricular septal defect with pulmonary stenosis (p 322) It is my belief that early in life these patients have a pulmonary resistance considerably lower than the systemic resistance As the child grows and as the right side of the heart hypertrophies the pulmonary obstruction becomes more and more significant and the predominantly left to right or balanced shunt through the ventricular defect becomes a dominantly right to left one A loud systolic murmur is always present at the lower left sternal border during infancy a diastolic rumble may be heard These patients do not have anoxic spells but dyspnea on exertion becomes more manifest through the years Squatting may not appear until five or six years of age or even later Cyanosis is minimal at rest but becomes noticeable on exercise and in cold weather Polycythemia is not present but the levels for hemoglobin and erythrocytes are high normal Sometimes with this minimal symptomatology adaptation is extremely difficult for these children For all practical purposes they look like everyone else and still they cannot do nearly as much as can the majority of their contemporaries Probably life expectancy should be placed between the second and fourth decades or even higher in certain cases The causes of death are the same as those mentioned in the discussion of the previous group

Complications

Among the complications of tetralogy of Fallot a number of rather important entities should be mentioned

Brain Abscess If signs and symptoms pointing to involvement of the central nervous system develop in a patient with tetralogy of Fallot or other congenital heart disease with a right to left shunt the diagnosis of a brain abscess should be thought of immediately These patients are usually more than two years of age and are polycythemic A minor febrile illness sore throat or upper respiratory infection occurs first About seven to ten days later headaches may develop vomiting occurs and there are personality changes and perhaps focal neurologic lesions with convulsions Fever may or may not be present The significance of papilledema may be hard to evaluate in a child with severe cyanotic disease Changes in the motor and sensory spheres may be

elicited and pathologic reflexes may be present A positive Kernigs sign may be noted Laboratory examination may reveal leukocytosis although brain abscess is one of the few instances in which a localized collection of pus may occur without an increase in the polymorphonuclear cell count Lumbar puncture usually shows some increase in cerebrospinal fluid protein content the cell count may or may not be increased and the sugar content is usually normal The electroencephalogram may indicate a localized process

Under these circumstances as stated before the diagnosis of brain abscess should be suspected and the advice and aid of a neurosurgical consultant should be sought It may be preferable to adopt the conservative attitude of careful observation or it may be better to proceed immediately by drilling burr holes over the right parieto occipital region the most common site of abscesses in this condition or wherever the electroencephalogram shows localized abnormalities [176] If the neurosurgical consultant prefers to watch the patient for a few hours or days evidences of acute meningitis may manifest themselves as the abscess starts leaking At this stage another lumbar puncture is indicated this usually reveals a high cell count and a low sugar content As soon as the diagnosis of brain abscess is made surgical evacuation is indicated with simultaneous antibiotic control of the infection Within the past five years at least three brain abscesses in patients with the tetralogy of Fallot have been operated on successfully by the neurosurgeons at The Childrens Medical Center in Boston

Why patients with tetralogy of Fallot are prone to the development of brain abscesses has not been satisfactorily explained so far It is possible that infected emboli from the systemic venous reservoir are carried across the ventricular defect and propelled directly through the carotid arteries up to the central nervous system It is also perfectly feasible that cerebral thrombi inherent in the polycythemia of these patients may become secondarily infected during a transient episode of bacteremia The organism recovered from the abscess is usually the alpha streptococcus although other etiologic agents such as *Hemophilus influenzae* *Staphylococcus aureus* gram negative and gram positive rods and *Pseudomonas aeruginosa* have all been encountered

Hemiplegia Hemiplegia without any evidence of a space occupying lesion of the central nervous system is probably due to cerebral thrombosis embolism or profound anoxia It usually occurs in infants less than two years of age with marked unsaturation There is no clear cut correlation between the hematocrit level and the occurrence of a cerebrovascular accident Sometimes its appearance follows a severe anoxic spell at other times it is noted immediately after a shunt operation It may be more common in hot weather with consequent dehydration or during febrile illnesses when the patient refuses to drink

Obviously this condition for which only palliative treatment and

physiotherapy are available should be carefully differentiated from a remediable brain abscess If the clinical picture and the cerebrospinal fluid findings are equivocal burr holes or even exploration may be indicated *It may be possible to prevent this type of cerebrovascular* accident by making certain that the patient has an adequate fluid intake under all circumstances and that any infections are promptly treated I am not convinced of the value of venesection in these patients

Relative Anemia and Polycythemia One of the adaptative mechanisms of the organism to low arterial oxygen saturation is the increase in oxygen carrying capacity (p 277) of the blood by the development of polycythemia and increased hemoglobin content of the individual red cells Indeed it may be demonstrated that cyanotic infants with low erythrocyte counts and low hemoglobin levels perform less adequately and have considerably more symptoms than if their hemoglobin and red *cell counts were raised to more adequate levels* The limiting factor of this mechanism is obviously the increasing viscosity of the blood It may be seen from Figure 293 that when the hematocrit reaches 65 to 70 per cent *further increase will entail a considerable rise in blood viscosity* resulting in impediment of blood flow sludging and even decreased delivery of oxygen to the tissues

On the basis of these observations a theory of optimal hematocrit for patients with cyanotic disease has been developed [97] We have seen patients with severe arterial unsaturation and hemoglobin levels of 10 to 13 gm per 100 ml of blood erythrocyte counts of 6 to 8 million and hematocrit levels of 40 per cent these children have a relative hypochromic anemia easily correctable by iron administration The resulting increase in hematocrit is usually accompanied by marked symptomatic improvement until the level reaches 70 to 75 per cent after which the symptoms observed at the low hemoglobin level recur

At present it is our policy to treat with iron all patients with tetralogy of Fallot who are hypochromic irrespective of the actual level *of erythrocytes We usually stop or decrease iron administration when* the hematocrit level reaches 75 per cent If when the patient is first encountered he has a hematocrit reading of 80 per cent or more and is *severely symptomatic venesection may be cautiously* attempted *If the* removal of small increments of blood results in a decrease in symptoms it may be repeated until the hematocrit level reaches 65 to 70 *per cent If* on the other hand symptoms do not improve or if they become aggravated by venesection the procedure should be abandoned immediately If the patient is relatively asymptomatic to begin with venesection is not recommended at *all* regardless of the level of the hematocrit

In summary I believe that patients with tetralogy of Fallot and appreciable arterial unsaturation probably function best with a hematocrit level of 55 to 75 per cent If the values are lower than this iron is

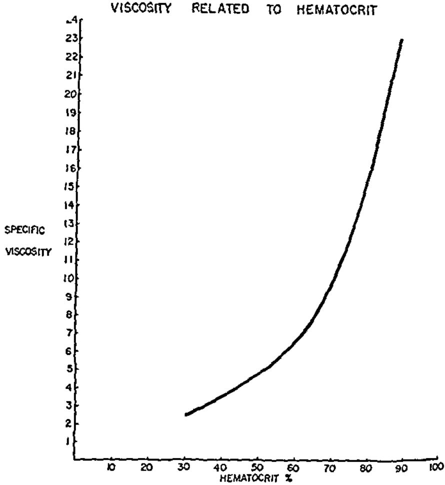


Figure 293 Correlation of hematocrit with the specific viscosity of the blood [9]

indicated if they are higher and the patient is severely symptomatic venesection may be attempted cautiously

Rheumatic Fever and Rheumatic Heart Disease Rheumatic fever and rheumatic heart disease are rare in patients with tetralogy of Fallot

Subacute Bacterial Endocarditis Subacute bacterial endocarditis represents a statistically significant danger for children with tetralogy of Fallot

Differential Diagnosis

Pulmonary Stenosis with Intact Ventricular Septum See page 449

Single Ventricle See page 340

Truncus Arteriosus See page 375

Tricuspid Atresia Only on the basis of the electrocardiographic

evidences of left ventricular hypertrophy can tricuspid atresia be differentiated from tetralogy of Fallot In every other respect they may be indistinguishable from each other

Aortic Atresia Aortic atresia should seldom be confused with the tetralogy of Fallot because of the large heart and deep cyanosis noted at birth

Complete Transposition Complete transposition of the great arteries with pulmonary stenosis presents a clinical picture which for all practical purposes may be indistinguishable from that in severe tetralogy of Fallot This should hardly be surprising in view of the fact that in both conditions pulmonary stenosis is present and the aorta receives right ventricular blood Clearly then the differences are quantitative rather than qualitative Clinical hints suggesting complete transposition of the great arteries may be found in the fact that these infants are usually grossly underdeveloped their hearts tend to be large their cyanosis is usually fairly severe and survival beyond the age of one year is not common Cardiac catheterization usually is not helpful in the differential diagnosis but angiocardiography in the sharp left lateral position revealing the aorta to communicate only with the right ventricle may be diagnostic of complete transposition of the great arteries

Levocardia with Abdominal Situs Inversus and Absent Spleen These patients if the examination is restricted to the cardiovascular system may look like typical patients with tetralogy of Fallot If however an abdominal situs inversus is discovered on physical examination or at fluoroscopy the suspicion of the absent spleen syndrome should be raised immediately In the comprehensive monograph of Ivemark[17] it is pointed out that this syndrome is almost never accompanied by a straightforward tetralogy of Fallot type of lesion but includes with a high statistical probability lesions such as atrioventricularis communis truncus arteriosus pulmonary atresia or transposition of the great arteries As expected the usual surgical procedures used for patients with the tetralogy of Fallot prove fatal to a large proportion of these children

Treatment

In 1945 Blalock and Taussig[6] reported the first results of their shunt" operation aimed at increasing the pulmonary blood flow by anastomosing a systemic artery (usually the subclavian) to the pulmonary artery Potts and his associates achieved the same purpose by establishing a direct connection between the aorta and the pulmonary artery[26] More recently Brock advocated directly relieving the stenosis of the right ventricular outflow tract Finally Lillehei and his associates[14] with the help of cross circulation and the Mayo Clinic group[104] with the aid of a pump oxygenator were able to create a bloodless field

in the heart and thus repair both the ventricular septum and the pulmonary stenosis in patients with tetralogy of Fallot

The desirability of the latter two procedures that is the complete repair of both major defects of tetralogy of Fallot depends entirely on the safety of the operation At the writing of this book the procedures still involve a 20 to 25 per cent mortality risk this seems too high a figure particularly since palliative operations with much lower mortality rates are available Nevertheless although today I probably should not recommend direct operation for a patient with uncomplicated tetralogy of Fallot there is little doubt that this will be the procedure of choice within a short time—that is as soon as the safety of the surgical approach is increased In the meantime the availability of complete repair colors significantly the indications for palliative operation Since direct repair is just "around the corner" a shunt procedure is not usually recommended unless the symptoms are really severe

By definition the three palliative procedures—Blalock Potts and Brock—do not aim to correct all the components of the tetralogy of Fallot They are designed simply to increase pulmonary flow and thus to increase the oxygen content of the mixed blood offered to the aorta As a result of successful operation there is a decrease in cyanosis and clubbing with a significant improvement in exercise tolerance

Of the three palliative procedures two are shunt" operations that is they establish a communication between the aorta and the pulmonary artery either directly (Potts) or via the subclavian artery (Blalock) In effect then another malformation is added to the defects of tetralogy of Fallot At The Childrens Medical Center most of our experience in this line has been with the Potts procedure Our surgical staff performs a left thoracotomy on all these patients and if the aortic arch is found to be on the left a Potts procedure is carried out Conversely if the arch is on the right a Blalock operation is performed using the left subclavian artery [151] The only deviation from this policy occurs in small infants in whom a Potts procedure is practically mandatory thus for a right aortic arch in an infant a right thoracotomy is performed

Our over all mortality with this approach has been 11 per cent In children less than three years old the mortality was 30 per cent while in patients more than three years old it was only 7 1 per cent Potts [5] reported on 500 unselected cases with a mortality of 15 per cent in the children less than three years old and 2 8 to 4 6 per cent in those more than three years of age Taussig and Bauersfeld[333] reported comparable mortality figures in their first 1000 cases

The operative results are dramatic Approximately 90 per cent of the survivors are able to lead relatively normal lives They are able to go to school and to enjoy noncompetitive sports they do not squat and their color is practically normal The remaining 10 per cent show moderate improvement or none at all

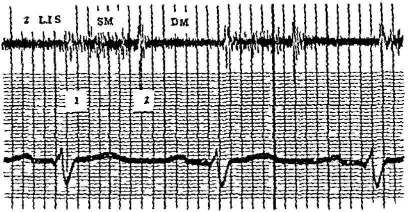


Figure 294 Continuous murmur in tetralogy of Fallot patient after successful Potts procedure

The measure of the success of the operation can often be judged best by the development of a continuous ductus like murmur (Fig 294) at the second interspace on the side of the operation Usually with the Potts procedure the murmur is audible within the first day after operation while with the Blalock operation the murmur may not be audible for a week or more Evidences of left ventricular hypertrophy become noticeable in the electrocardiogram (Figs 295 296) left ventricular and left atrial enlargement are commonly seen by x ray (Fig 297) Pulmonary vascular engorgement of slight to moderate degree may be visible in the postoperative film

How long after a shunt procedure this clinical improvement lasts is impossible to tell It may be said, however that few of the people (less than 1 per cent) whose operation was initially successful need reoperation if a child shows definite improvement within the first six postoperative months the results are still good as many as ten years later The anastomosis if adequate within the first six months rarely if ever closes as far as we know at the present time If the anastomosis closes—if the continuous murmur disappears—it usually was not satisfactory in the beginning

If the operative shunt (usually the Potts type) is too large the patient may go into congestive failure within the first week or two after operation This is evidenced by a hyperdynamic cardiac impulse pulmonary rales systemic congestion appearance of apical diastolic murmur wide pulse pressure and marked cardiac enlargement In most instances this can be remedied by vigorous anticongestive measures pursued for six to eighteen months Eventually perhaps through the development of some degree of pulmonary vascular obstruction the circulation adjusts itself and within one or two years the patient begins to show the full benefits of the operation (Fig 298)

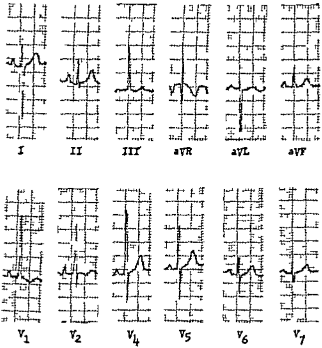


Figure 295 Electrocardiogram of patient with tetralogy of Fallot

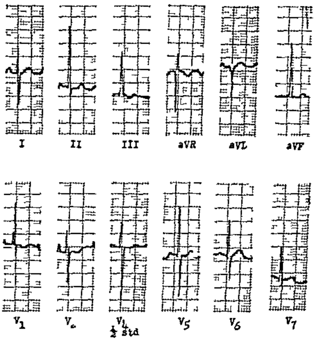


Figure 296 Electrocardiogram of the same patient as in Figure 295 after a Potts procedure Note the increase in left ventricular potentials in leads aVF and V7 Also the increased voltages in leads II and III

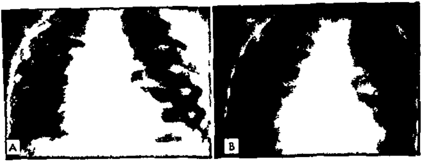


Figure 297 Posteroanterior radiograms of a patient before (*A*) and after (*B*) a Potts procedure *A* Note ischemic lung fields and a small heart *B* Note pulmonary vascular engorgement particularly in the region of the left pulmonary artery and some cardiac enlargement.

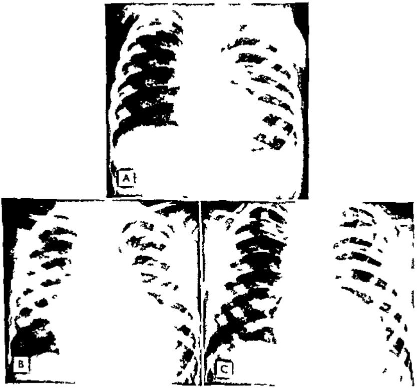


Figure 298 *A* Posteroanterior view of typical tetralogy of Fallot *B* Posteroanterior view of the heart of the same patient after operation, showing marked cardiac enlargement and pulmonary vascular engorgement, three months later C Disappearance of pulmonary congestion and diminution of heart size after adequate anti congestive treatment

Of course if evidences of congestive failure appear postoperatively in a patient thought to have tetralogy of Fallot one must be certain that the preoperative diagnosis was not faulty and that the child is not suffering from pure pulmonary stenosis or a transposition of the great arteries neither of which lends itself to symptomatic relief by means of this technique If the diagnosis was wrong obviously anticongestive measures will not satisfactorily solve the problem the anastomosis may have to be closed and an appropriate operation if technically possible, performed

A word may be said here about the reasons why pure pulmonary stenosis and transposition of the great arteries do not respond well to a shunt procedure In transposition of the great arteries the answer is simple Diminished pulmonary blood flow is *not* the principal cause of cyanosis in these patients furthermore establishing an aortopulmonary anastomosis does not necessarily establish the type of shunt that is most useful under the given circumstances Even if an aortopulmonary shunt is needed in transposition of the great arteries with pulmonary stenosis without an appropriate sized atrial septal defect such a shunt may have disastrous results

Why patients with pulmonary stenosis of the isolated type respond poorly or even go into congestive failure after a shunt procedure has become clear only relatively recently If the patient with this anomaly is not cyanotic then obviously a shunt operation recirculating fully oxygenated blood through the lungs will not accomplish anything beneficial If the patient s blood is unsaturated because of a right to left atrial shunt then the cyanosis may be helped by the shunt operation but it does not remedy the more serious underlying problem As mentioned before in patients with pure pulmonary stenosis a shunt from right to left will not occur until right atrial pressure is higher than left atrial pressure that is until the right ventricular diastolic pressure is elevated This on the other hand means right ventricular failure In this situation then the basic difficulty is right ventricular failure a condition not remediable at all by establishing an aortopulmonary shunt (This is in marked contrast to tetralogy of Fallot in which congestive failure does not occur in childhood) Consequently patients with pure pulmonary stenosis and a right to left shunt go into congestive failure after a Potts procedure principally because they would have done so anyhow Another added reason for the development of failure postoperatively is the fact that as mentioned earlier the increased left ventricular flow work may easily lead to left ventricular failure in any patient after a shunt operation Left ventricular failure then leads to left atrial and pulmonary capillary hypertension This on the other hand leads to pulmonary arterial hypertension and increased right ventricular work

The complications of tetralogy of Fallot enumerated on page 465

apparently do not occur any more frequently in patients after a shunt procedure than before

On the whole the shunt operation once its essentially palliative nature is appreciated is a satisfactory procedure which gives maximal symptomatic relief with minimal risk However it does increase significantly the work of the heart, and after a number of years this will probably cause congestive failure in many if not all patients operated on

The search for a more physiologic approach to the problem of pulmonary stenosis with ventricular defect and right to left shunt led Brock to consider the use of his valvotomy technique in these patients At the time this was first mentioned it sounded like a most attractive idea and to some people it still does Although even the most ardent supporters of the procedure have to admit that the operation fails to cope with the ventricular septal defect, even its most vociferous enemies must admit that the Brock procedure at least relieves one condition instead of adding another anomaly to those already existing The operative results in the most successful cases have been magnificent. Patients operated on by Brock and by others in this country have shown more striking clinical improvement than those with the best results from the shunt operation Without the use of the stethoscope one could barely distinguish these children from normal ones There are a number of difficulties to be considered, however

First the mortality figures even in Brocks hands are approximately 12 or 13 per cent,[53] and few if any of these patients have been small infants Thus one is confronted with a palliative procedure which entails a considerably higher risk than the shunt operation Second, Brocks experience of finding pure valvular stenosis in 30 per cent of his cases and combined valvular and infundibular stenosis in another 15 per cent, does not correspond to our experience or to the experience of most American observers [18, 63] Therefore in most instances we would have to use the less favorable mortality figures relating to infundibular resection rather than the more favorable ones applying to valvotomy Third the postoperative course is stormy in many of the patients Significant cardiac enlargement pericardial effusion congestive failure (Fig 299) and pulmonary regurgitation (Fig 300) all are rather common complications of the Brock procedure in patients who have pulmonary stenosis with a ventricular septal defect Many though not all of these patients recover but are left with a loud murmur of pulmonary regurgitation and a considerably enlarged heart Finally our surgeons at The Childrens Medical Center believe that eventual complete repair of both the infundibular stenosis and the ventricular septal defect may be considerably easier if the myocardium has not previously been damaged by a transventricular incision

All these considerations lead us to believe that in most cases of

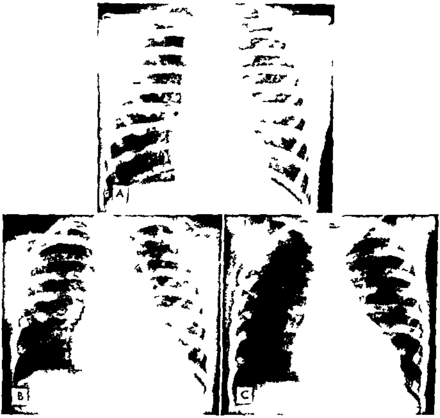


Figure 299 Posteroanterior radiograms of patient with a tetralogy of Fallot undergoing a Brock procedure A Preoperative radiogram B Immediately postoperatively C Three months later Note the marked cardiac enlargement appearing immediately after operation which recedes to a certain extent in the last film

tetralogy of Fallot at this time a shunt procedure may be preferable to the Brock approach We are cognizant of the fact that both these operations are only temporary expedients once the palliative nature of the operation is recognized we prefer the approach carrying the least risk and offering the least chance of jeopardizing further operation on the heart

The indications for palliative surgery in patients with tetralogy of Fallot are twofold In one group of patients—usually small infants with severe anoxic spells—it may be a lifesaving measure In these situations prompt operation is recommended if the spells either occur so frequently (several times a day) or are so severe (loss of consciousness convulsions) that the infant s life is endangered As mentioned earlier the mortality in this group is high but there is really no choice but to go ahead with the operation

The second group of patients in whom a shunt procedure should be considered are those whose dyspnea on exertion is so severe that they

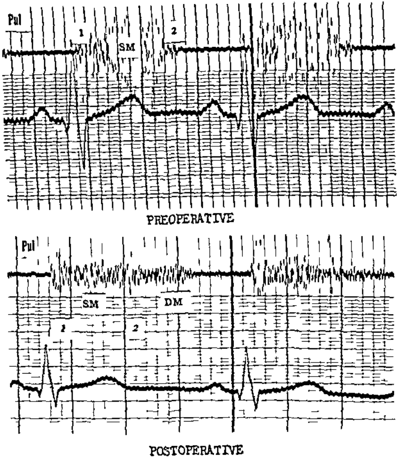


Figure 300 Preoperative and postoperative phonocardiograms in a patient with tetralogy of Fallot who has undergone a Brock procedure Note the appearance of long diastolic murmur in the postoperative record

are unable to lead a relatively contented and happy life Severe dyspnea on exertion is rather well tolerated by toddlers who play at home and can set their own pace however as mentioned before an entirely different situation arises when they enter school One further point to be considered in appraising the group is the course of the disease Has there been recent deterioration or improvement in the patient s condition or has it remained unchanged? If on the whole the patient is better off this year than last year or the year before we would not advise operation at present On the other hand if the symptoms within the past months or year have been bad or have been getting worse operation is recommended In this group of children the optimal age for operation is probably between four and fifteen years

Preoperative catheterization or angiocardiography is not indicated

if the clinical profile is classical The hematocrit level before operation should be approximately 50 to 60 per cent venesection or packed cell transfusion helps to achieve this Digitalis quinidine or procaine amide is not given preoperatively unless specifically indicated The postoperative course is usually uneventful

Medical management is important in the handling of these children In perhaps no other part of pediatrics is the art of medicine put to a more strenuous test than in the skillful handling of these patients and their parents Sometimes it is extremely hard to keep up the parents courage and teach them to accept the situation as one with which they may have to live for a number of years Only slightly less difficult is the decision when to advise operation one must either take the risk of a number of fairly severe spells or be ready to face the considerable dangers of operation in young infants All these problems the physician must solve to the best of his ability and with considerable humbleness knowing that his decisions unfortunately will not always be correct One small practical point should be mentioned if these children are admitted to a hospital ward for study or operation the staff should be forewarned that "routine" blood tests and other laboratory tests should be taken only with the utmost caution and sometimes only under sedation Major spells and consequent death have been caused by unskilled attempts at venipuncture in a small child with a tendency toward anoxic spells

SUMMARY

The large majority of patients more than two years of age with cyanotic congenital heart disease have pulmonary stenosis with a right to left shunt through a ventricular defect—that is the tetralogy of Fallot The clinical profile is characterized by a small heart a stenotic murmur at the left sternal border a diminished pulmonary second sound poorly vascularized lung fields and right ventricular hypertrophy The principal handicaps resulting from this defect are anoxic spells during infancy and severe dyspnea on exertion throughout childhood and adult life A shunt operation is a lifesaving measure as far as the spells are concerned and it affords symptomatic relief for the exercise intolerance Eventually complete repair of the ventricular defect and the pulmonary stenosis is desirable

TETRALOGY OF FALLOT WITH UNILATERAL PULMONARY ATRESIA

It has been pointed out in the previous section that the shunt operation is in my opinion the procedure of choice at this time for correction of tetralogy of Fallot It is implicit in the operative technique that the successful accomplishment of an aortopulmonary shunt depends to a large extent on the availability of a pulmonary artery through which the systemic blood can be directed to the lungs and on the presence

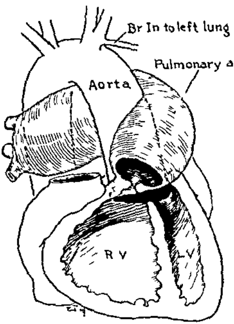


Figure 301 Diagrammatic presentation of tetralogy of Fallot with atresia of the left main pulmonary artery and marked dilatation of the right main pulmonary artery [57]

of a second vessel to maintain the pulmonary circulation during the period of temporary occlusion of the first vessel A rather rare variant of tetralogy of Fallot in which only one main pulmonary artery is present will be discussed here briefly because of the specific difficulties that this situation presents during operation

INCIDENCE

This apparently is a rare condition—we have found only five such cases during the past ten years in our clinic [57]

ANATOMY

The anatomic situation is similar to that found in the classical tetralogy of Fallot Pulmonary stenosis (usually valvular) is associated with a ventricular defect overriding aorta and right ventricular hyper trophy The only distinguishing feature is that one pulmonary artery (usually the right) shows considerable poststenotic dilatation while the other is completely absent (Fig 301)

PHYSIOLOGY

Hemodynamically the situation is also practically identical with that found in tetralogy of Fallot except that the circulation to the lung which is not supplied by a pulmonary artery is exclusively through collateral channels

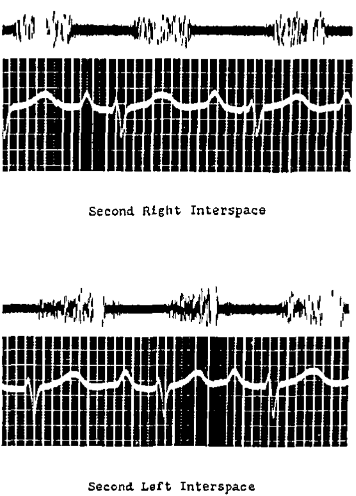


Figure 302 Phonocardiogram of patient with atresia of the left main pulmonary artery Note the loud murmur at the second right interspace with considerably fainter murmur at the second left interspace [237]

Clinical Picture

In terms of symptomatology the clinical picture cannot be distinguished from that of straightforward pulmonary stenosis with ventricular defect and a right to left shunt of rather severe degree

On physical examination the only distinguishing feature in these patients is that the stenotic murmur propagates almost exclusively to the side where the large main pulmonary artery is This results in a murmur at the second right interspace (Fig 302) in patients with levocardia and at the second left interspace in those with dextrocardia

The electrocardiogram reveals the usual pattern of right ventricular hypertrophy

The roentgenologic picture is the only truly diagnostic feature of



Figure 303 Posteroanterior radiogram of patient with atresia of the left main pulmonary artery Note the ischemic left lung field with plethoric right lung field

this condition these patients show marked diminution of the pulmonary vasculature and a small lung volume on one side and a dilated main pulmonary artery with pulmonary vascular engorgement and even occasionally hilar dance on the other (Fig 303)

At cardiac catheterization valvular pulmonary stenosis with right ventricular hypertension in the systemic range is encountered

Angiocardiography is the most specific diagnostic tool revealing the overriding aorta a large right main pulmonary artery and no opacification of the left main pulmonary artery (Fig 304)

Course and Prognosis

The expected course of this entity is difficult to outline with any accuracy because of the meager number of patients available Three of our five patients live severely restricted lives and are five to ten years of age at present One infant experienced severe pulmonary difficulties because of the compression of the right main bronchus by the aneurysmally dilated right main pulmonary artery this infant died after an attempt at surgical correction A fifth patient with extremely severe symptoms was successfully operated on

Differential Diagnosis

Tetralogy of Fallot Tetralogy of Fallot may be distinguished from this entity by the uniformly ischemic lung field and by the transmission



Figure 304 Angiocardiogram with diagrammatic explanation in a patient with atresia of the left main pulmonary artery [94]

of the systolic murmur to the left side of the chest if the heart is in the normal position

Pulmonary Vascular Obstruction with Ventricular Defect and Right to Left Shunt Pulmonary vascular obstruction with ventricular defect and right to left shunt may be confused with tetralogy of Fallot with unilateral pulmonary atresia because of the pulmonary plethora in

one hemithorax The presence of the stenotic murmur and the uneven distribution of the vasculature suggest the correct diagnosis rather easily

Hemitruncus Hemitruncus with a large pulmonary artery to one lung and no pulmonary artery to the other cannot be differentiated from this condition except by means of an angiocardiogram in the correct position

Treatment

Because of the technical difficulties outlined in the introductory paragraph operation probably ought not to be recommended for the time being unless the symptoms are well nigh intolerable In that case a right Blalock anostomosis to the large right main pulmonary artery is recommended The operative risks are high

Summary

Patients who have a variant of tetralogy of Fallot with unilateral atresia of one pulmonary artery and poststenotic dilatation of the other may be recognized by the peculiar transmission of the murmur and the uneven distribution of the vascular pattern as shown on the roent genogram Since operation is fraught with more than average risk it should not be recommended unless it is absolutely necessary If a shunt procedure still seems desirable the anastomosis should be effected to the large pulmonary artery rather than attempting to find a vessel in the completely undervascularized lung field

IDIOPATHIC PULMONARY REGURGITATION

Incidence

Whereas incompetence of the pulmonary valve in patients with pulmonary vascular obstruction (idiopathic or associated with congeni tal heart disease or mitral stenosis) is a fairly common occurrence pulmonary regurgitation as an isolated phenomenon is rare One such case has been described by Kjellberg and his associates[191] We have seen three such patients at The Children s Medical Center in Boston

Anatomy

To the best of my knowledge no anatomic studies are available in any patients with this syndrome In all likelihood the condition is con genital

Physiology

Patients with idiopathic pulmonary regurgitation are characterized by the presence of a loud early diastolic murmur in the pulmonary area and low diastolic pressure in the pulmonary artery without any other hemodynamic abnormality Physiologically the results of even maximal

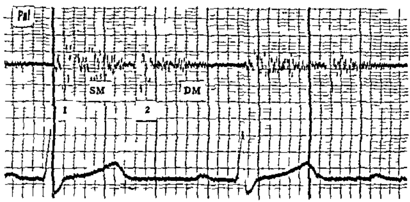


Figure 305 Phonocardiogram of patient with idiopathic pulmonary regurgitation

pulmonary insufficiency are slight [1 1] the normally low diastolic pressure in the pulmonary artery precludes the regurgitation of large volumes of blood during ventricular diastole

Clinical Picture

No symptoms attributable to heart disease are present On physical examination the most striking finding is a loud early diastolic murmur even accompanied by a diastolic thrill at the second to fourth left inter space with a decrescendo configuration (Fig 305) A systolic murmur of lower intensity and frequency may also be heard in the same area, however this lacks the typical characteristics of a stenotic murmur The heart sounds are normal interestingly enough the pulmonary second sound is of only average intensity and splitting The rest of the physical *examination is entirely negative Most important the pulse pressure* is well within normal limits

Roentgenologically no increase in the transverse diameter of the heart is noted The main segment of the pulmonary artery is prominent and pulsates vigorously (Fig 306) The pulmonary vasculature is well within normal limits

The electrocardiogram is normal

Cardiac catheterization shows normal hemodynamics with the exception of a low pulmonary arterial diastolic pressure

Angiocardiographic studies have not been performed on any of our patients

Course and Prognosis

On the basis of limited experience I believe that this condition is probably compatible with normal life expectancy and a fully active life

Differential Diagnosis

Aortic Regurgitation Sometimes it is difficult to distinguish pulmonary from aortic regurgitation on the basis of the localization and the



Figure 306 Posteroanterior radiogram of patient with idiopathic pulmonary regurgitation Note the relatively normal sized heart with markedly prominent main pulmonary artery but normal pulmonary vasculature

character of the murmur alone The most important points in differential diagnosis are the wide pulse pressure and the history of rheumatic fever in patients with aortic regurgitation

Patent Ductus Arteriosus or Ventricular Septal Defect with Aortic Regurgitation Patent ductus arteriosus or ventricular septal defect with aortic regurgitation are relatively easily distinguished from idiopathic pulmonary regurgitation by the presence of pulmonary vascular engorgement in the roentgenogram Occasionally cardiac catheterization is necessary to exclude the presence of a small left to right shunt

Pulmonary Vascular Obstruction Perhaps the most common cause of pulmonary insufficiency pulmonary vascular obstruction is distinguished from idiopathic pulmonary regurgitation on the basis of evidences of pulmonary arterial hypertension such as a ringing pulmonary second sound and evidences of right ventricular hypertrophy in the electrocardiogram

Valvular Pulmonary Stenosis Valvular pulmonary stenosis of maximal degree may be accompanied by a murmur suggestive of pulmonary regurgitation As pointed out earlier actually this is only a continuation of the systolic murmur beyond the aortic component of the second sound, a phenomenon relatively easily demonstrated phonocardiographically Furthermore the systolic murmur is always louder and stenotic in character

Treatment

No treatment is necessary for idiopathic pulmonary regurgitation beyond reassurance to the parents and the child

Summary

An early diastolic murmur at the pulmonary area accompanied by roentgenologic evidence of a large main pulmonary artery segment and perfectly normal hemodynamics characterizes the syndrome of idiopathic pulmonary regurgitation The condition is innocuous and requires no treatment

TRICUSPID ATRESIA

In tricuspid atresia no direct communication exists between the right atrium and the right ventricle The right atrial blood is shunted through a foramen ovale or an atrial defect into the left atrium where complete mixture with the pulmonary venous blood occurs

Incidence

Tricuspid atresia has been regarded as one of the rarest congenital cardiac diseases but it must be considered in the differential diagnosis of every patient with cyanotic congenital heart disease Brown[56] in the latest edition of his book mentioned fifty cases collected from the literature Abbott[1] reported sixteen in her series We have seen twenty three proved cases at The Children's Medical Center in Boston within the past ten years and are following up at least twenty five more patients in whom this diagnosis is based on clinical observations alone Thus in my opinion this anomaly may be more common than was hitherto suspected Campbell[7] stated that patients with tricuspid atresia constitute about 5 per cent of the patients with cyanotic congenital heart disease

Anatomy

On the basis of the anatomic situation in the ventricles Edwards and his co workers[10] distinguished four types of tricuspid atresia depending on whether or not the great arteries are transposed whether a defect in the ventricular septum exists and whether there is stenosis of the pulmonary artery (Fig 307)

Embryologically the defect probably occurs about the fourth or fifth intrauterine week and is due to an incomplete shift of the common atrioventricular canal so that the ventricular septum growing up from below fails to divide it evenly The lesions associated with tricuspid atresia such as atrial defect, patent foramen ovale patent ductus arteriosus or hypoplastic pulmonary artery probably result from the altered hemodynamics caused by the tricuspid atresia

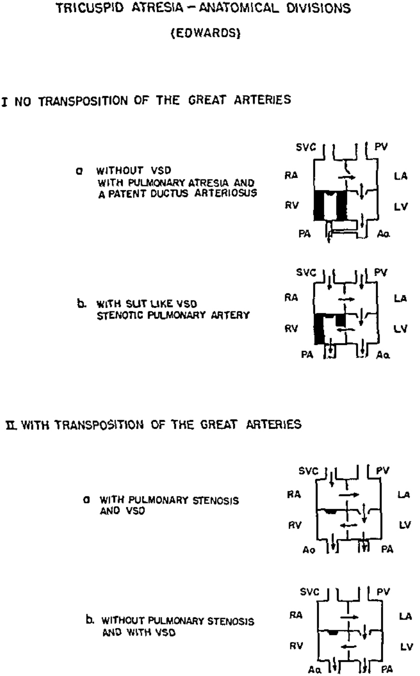


Figure 307 Schematic presentation of the varieties of tricuspid atresia (After Edwards [97])

PHYSIOLOGY

The physiologic consequences of tricuspid atresia are manifold[42] The first and most important result is the presence of arterial unsaturation due to a complete mixing of systemic venous and pulmonary venous blood in the left atrium The degree of unsaturation depends on the ratio of pulmonary venous to systemic venous return within this common left atrial pool this in turn is dependent entirely on the size of the pulmonary blood flow The larger the pulmonary blood flow the less marked is the cyanosis thus patients of types Ia Ib and IIb of Edwards classification have marked cyanosis (Fig 307) On the other hand patients of type IIb, with large pulmonary blood flow do not have to show definite cyanosis at all

A second and equally important physiologic consequence of tricuspid atresia is the underdevelopment of the right ventricle in types Ia and Ib as a result of the inadequate blood flow through it The third physiologic handicap is the elevated right atrial pressure necessary to force the caval blood over to the left atrium this in turn results in increased systemic venous pressure and eventually congestive failure This then results in the paradoxical situation of right atrial hypertension without concomitant right ventricular hypertension

CLINICAL PICTURE

The clinical picture to be described is based principally on the findings in our twenty three proved cases of tricuspid atresia

These children are all sick in early infancy Severe cyanosis is the dominant symptom we have seen only two patients in whom little or no cyanosis was noted In addition dyspnea fatigue anoxic spells and of great importance evidences of right sided failure are observed Squatting is rare

At physical examination the majority of the infants are markedly undernourished and poorly developed cyanosis and clubbing are readily apparent Left sided chest prominence is absent the thorax either is symmetrical or may show a slight bulge on the right side of the sternum A thrill is palpable in only about a fourth of the patients Considerable hepatomegaly occasionally with presystolic pulsations is present in more than half Pulmonary rales and evidences of left sided failure are uniformly absent At auscultation the heart sounds are clear and narrow at the lower left sternal border Pulmonary closure is never accentuated as a matter of fact in most instances it is diminished in intensity A grade 3 to 4 harsh blowing systolic murmur is audible at the lower left sternal border in more than half of the patients In the others probably the sickest ones only faint murmurs or none at all are audible Interestingly enough no atrial sound or diastolic murmur is present

The electrocardiogram is always abnormal for the age of the patient Left ventricular hypertrophy (or left ventricular dominance in young

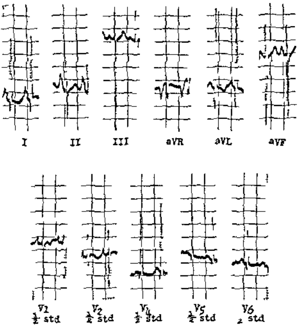


Figure 308 Electrocardiogram of patient with tricuspid atresia Note left axis deviation left ventricular hypertrophy and P pulmonale

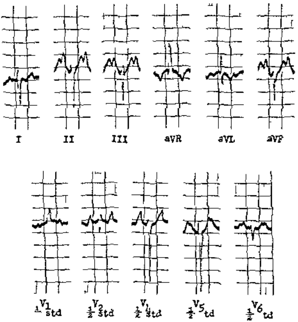


Figure 309 Electrocardiogram of patient with tricuspid atresia showing extreme right axis deviation with right ventricular hypertrophy in chest leads

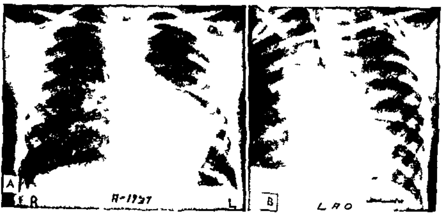


Figure 310 The classic radiological picture of tricuspid atresia in the postero-anterior and the left anterior oblique views Note the absent right border in the postero-anterior view and the cut off anterior border in the left anterior oblique view

infants) or P pulmonale is noted in almost every instance, frequently both features are present in the same patient (Fig 308) Left axis deviation in the standard limb leads is common I have seen patients with tricuspid atresia who had right axis deviation with right ventricular hypertrophy chest leads (Fig 309)

The roentgenologic findings have been summarized by Wittenborg and his associates [359] The classic picture (Fig 310) characterized by the straight right border with a concave pulmonary artery segment in the anteroposterior view and the straight anterior border in the left anterior oblique projection is certainly not the only radiologic configuration encountered in this disease Depending on the size of the interatrial communication and thus the size of the right atrium either the classic picture or a *coeur en sabot* configuration (Fig 311), or even the picture of dextrocardia (Fig 312) may be encountered Cardiac enlargement is always present and may vary from minimal to marked degree The pulmonary vasculature is greatly diminished except in the rare patient with complete transposition of the great arteries The main pulmonary artery segment is small or absent The aortic arch is on the left in 90 per cent of the cases No clear cut differentiation between right and left ventricular enlargement is possible by radiologic means Astley and his associates[17] described enlargement of the left atrium in the right anterior oblique and the posteroanterior view In the latter projection this gives rise to a "square heart."

No large series of patients have undergone cardiac catheterization therefore few physiologic data are available in this condition The meager observations we have made (Fig 313) indicate (1) There is no increase in oxygen content in the right atrium The catheter cannot be passed into the right ventricle but enters with relative ease the left atrium and



Figure 311 *Coeur en sabot* configuration in a patient with tricuspid atresia

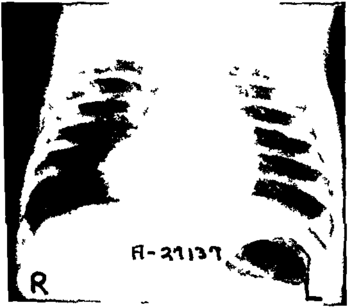


Figure 312 Dextrocardia like picture in patient with tricuspid atresia

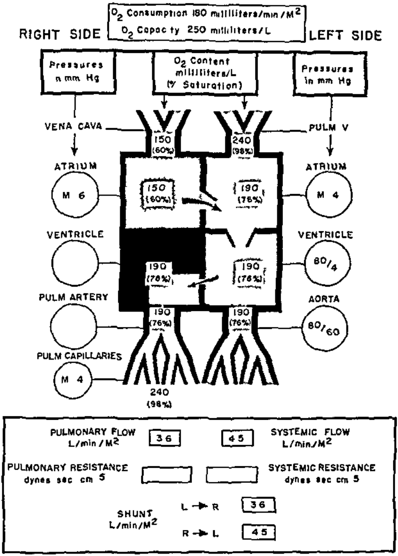


Figure 313 Schematic representation of cardiac catheterization findings in patient with tricuspid atresia

left ventricle Complete mixing of blood occurs in the left ventricle with oxygen saturation identical to that in the systemic artery (2) Right atrial pressure is elevated and a gradient may be demonstrated between the right atrial mean and left atrial mean systolic pressures (Fig 314) Often giant A waves are present in the right atrial pressure tracing The right ventricle and pulmonary artery may not be entered (3) Arterial saturation is low (4) Systemic flow is appreciably greater than pulmonary flow No accurate calculation of the respective resistances is possible

Angiocardiography if properly executed preferably by the Swedish

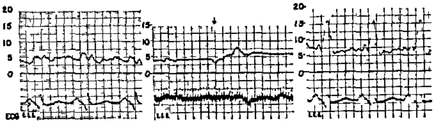


Figure 314 Withdrawal of catheter from left to right atrium Note the much taller A waves in the right atrial pressure tracing than in the left atrial record. Also note in the middle column the increase in mean pressure when entering right atrium from left atrium.

selective technique[191] is likely to furnish complete evidence for diag nosis Single plane angiograms are likely to be misleading sometimes it is impossible to tell from a single view whether the right to left passage occurs at the atrial or the ventricular level Optimally however a single plane angiogram may show clearly the passage of the dye from the right atrium to the left atrium to the left ventricle and the aorta The pul monary artery fills late and not distinctly A small unopacified area where the right ventricle usually is found may be noted (Fig 315)

Course and Prognosis

Cyanosis dyspnea and poor development are invariably noted in children less than six months old and in most cases even at birth Death usually occurs in congestive failure or in a severe anoxic spell within the first six to twelve months of life A minority of the patients usually those having a large atrial or ventricular defect may survive without operation until their third or even sixth birthday At present we have under observation a thirteen year old boy with tricuspid atresia deep cyanosis and severe limitations this is extremely rare however Patients with tricuspid atresia may experience the usual complications of cyanotic congenital heart disease that is hemiplegia anemia or polycythemia and subacute bacterial endocarditis However we have not yet encountered a patient with tricuspid atresia and brain abscess Obviously these pa tients do not live long enough to have rheumatic fever

Differential Diagnosis

The differential diagnosis of tricuspid atresia involves the cyanotic group of congenital heart diseases in which there may be electrocardio graphic evidences of left ventricular hypertrophy There are really only three conditions to be mentioned single ventricle (p 333) truncus arteriosus (p 370) and anomalous drainage of the entire caval system (p 528)

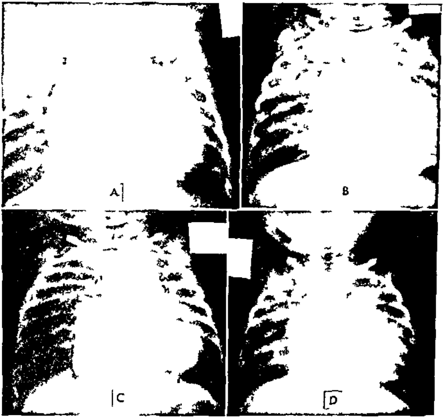


Figure 315 Angiocardiograms in patient with tricuspid atresia *A* The plain film without dye *B* Opacified right atrium with entrance of the dye into left atrium *C* Simultaneous opacification of right atrium left atrium and left ventricle *D* Dye in left atrium and left ventricle and aorta

Treatment

The treatment of patients with tricuspid atresia is surgical However no truly corrective procedure is available at present even in institutions equipped to perform open heart surgery Only palliative procedures—Blalock's or Potts—aimed at increasing the pulmonary blood flow have been performed on these patients and these have been only moderately successful Our own experience on a small group of patients indicates an almost 50 per cent operative mortality and only moderate improvement in the survivors Some of the patients (usually small babies) do not survive the operative procedure and others die some weeks later in congestive failure

These mortality figures as well as the symptomatic relief obtained in the survivors can be improved somewhat if at operation in addition to an aortopulmonary shunt the atrial defect is also enlarged through a left atrial approach This may increase the cardiac output and reduce

right atrial pressure Some of our patients with the best postoperative results have had this combined procedure and are alive at eight to twelve years of age

SUMMARY

Patients with cyanotic congenital heart disease whose electrocardiograms indicate left ventricular hypertrophy with P pulmonale have tricuspid atresia This is a severe anomaly causing cyanosis at birth and death usually within the first year of life Palliative surgery with a high mortality is the only available treatment

DOWNWARD DISPLACEMENT OF THE TRICUSPID VALVE (EBSTEIN S ANOMALY)

INCIDENCE

Ebstein s malformation is rare it was first described about 100 years ago Maude Abbott[1] did not mention any instance among her 1000 cases of congenital heart disease We have seen five patients with this anomaly in our clinic within the past ten years The total number of cases reported in the literature is certainly less than fifty

ANATOMY

In Ebstein s disease the posterior and frequently the septal leaflet of the tricuspid valve is not attached in a normal fashion to the annulus fibrosus but rather to the ventricular wall near the apex The anterior leaflet is conventionally fixed to the annulus fibrosus This anatomic constellation results in the upper part of the right ventricle being incorporated into the right atrium Thus the portion of the heart above the apex of the tricuspid valve is huge and can be divided into a superior (atrial) and inferior (ventricular) portion the border between the two is formed by the annulus fibrosus to which only the anterior leaflet is attached The ventricular cavity is small and consists only of the apical portion and the outflow tract of the right ventricle The walls of both the right atrium and the right ventricle are thin and flabby The infundibulum of the right ventricle is usually thick The tricuspid valve is not stenotic but may show a considerable amount of regurgitation Either the foramen ovale is open or an atrial septal defect is present

The embryology of this malformation has not been clearly explained so far

PHYSIOLOGY

I have no clear cut idea through what physiologic mechanisms the anatomic abnormality known as Ebstein s disease gives rise to the clinical picture to be described It is difficult to imagine why hypoplasia of the right ventricle alone should result in the characteristic clinical profile

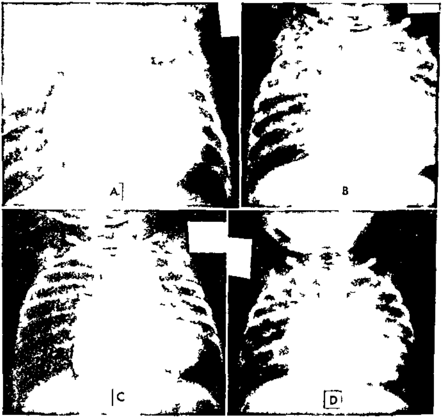


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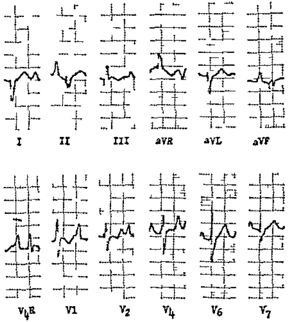


Figure 317 Electrocardiogram of patient with Ebstein s disease Note the marked P pulmonale with right axis deviation, incomplete right bundle branch block and questionable if any right ventricular hypertrophy

At auscultation a characteristic triple or quadruple rhythm at a slow rate and without clear cut gallop quality is the most striking feature phonocardiographic analysis (Fig 316) reveals the components to be the late first sound the widely split second sound and the loud fourth sound The pulmonary closure is diminished The systolic murmur may be vague and "scratchy" in quality or may assume a stenotic character if a gradient across the pulmonary valve exists The fourth heart sound is usually followed by a distinct presystolic murmur and occasionally is preceded by a mid diastolic rumble These auscultatory phenomena are pathognomonic of the condition and I have observed them in patients of varying ages from infancy to adulthood

The electrocardiogram is also typical (Fig 317) Large P pulmonale is clearly evident in most tracings Conduction disturbances (prolonged P R interval and incomplete or complete right bundle branch block) are the rule No right ventricular hypertrophy is noted after early infancy Left chest leads show average potentials Figure 318 demonstrates the change in one patient with characteristic right ventricular hypertrophy in early infancy and marked right bundle branch block without clear cut right ventricular hypertrophy four years later Arrhythmias such as premature beats and paroxysmal atrial tachycardia are common

The roentgenologic picture is also characteristic It consists of ex

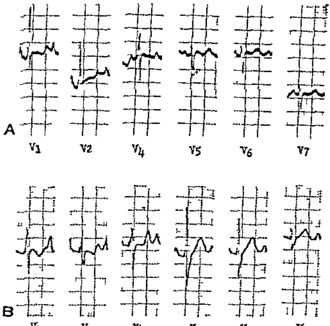


Figure 318 The development of the electrocardiogram in a patient with Ebstein's anomaly *A* Right ventricular hypertrophy in a child four months old without significant P pulmonale *B* Marked P pulmonale and disappearance of right ventricular hypertrophy and appearance of incomplete right bundle branch block particularly in lead V.

treme cardiac enlargement, involving principally the right atrium with diminished pulmonary vasculature (Fig 319) The pulmonary artery segment is hypoplastic At fluoroscopy the cardiac beat is feeble The entire silhouette of the heart and its activity resembles that in cases of a large pericardial effusion or pure pulmonary stenosis in congestive failure

Cardiac catheterization is a dangerous procedure in patients with Ebstein's anomaly because of the appearance of arrhythmias one of our patients with this anomaly died during catheterization It is because of this characteristic of the disease plus its extreme rarity that physiologic data are so sparse Observations on two of our patients and the data from the literature [3, 341] indicate the following characteristic catheter findings (Fig 320) (1) There is usually no significant increase in oxygen content throughout the right heart chambers and the pulmonary artery only rarely is a small left to right shunt at the atrial level noted (2) Right atrial pressure is high and the tracing suggests tricuspid regurgitation Right ventricular systolic pressure is usually normal end diastolic pressure is elevated Sometimes it is difficult to differentiate between right atrial and right ventricular pressures the contours of the two tracings may be similar differing only in the height of the systolic

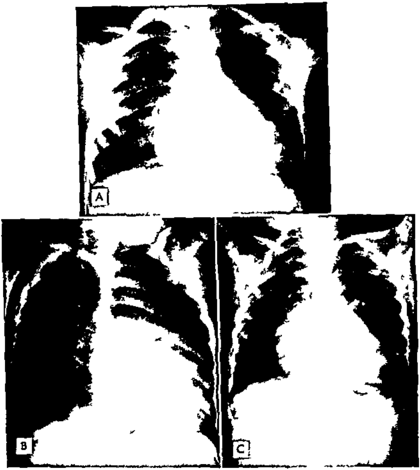


Figure 319 Posteroanterior and oblique radiograms of patient with Ebstein's disease Note the marked cardiac enlargement involving principally the right atrium and the diminished pulmonary vasculature

peak (Fig 321) (3) Pulmonary capillary pressure is normal as is the brachial arterial pressure (4) *Slight or moderate arterial unsaturation* is noted (5) Cardiac output is low normal Systemic flow is slightly larger than pulmonary flow (6) The catheter may easily coil up in the huge right atrial cavity

Angiocardiography is just as dangerous a procedure as cardiac catheterization in patients with Ebstein's disease One of our patients died thirty six hours after angiocardiography The information obtained by this examination is minimal indeed because of the stagnation of the dye *in the huge right atrium Consequently angiocardiography is not only dangerous but not particularly helpful In contrast cardiac catheterization although carrying a high risk furnishes the cardiologist with* an accurate diagnosis if it is successfully completed

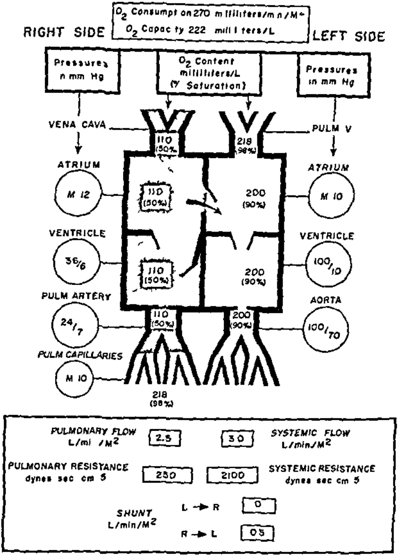


Figure 320 Diagrammatic representation of findings at cardiac catheterization in patient with Ebstein's anomaly [101]

Course and Prognosis

Because of the relatively few reported cases no accurate appraisal of the life history of these patients is possible at present It may be stated however that symptoms usually start in infancy or early childhood and are slowly progressive Death usually occurs in the second or third decade of life One patient has been reported to have reached sixty one years of age [109] The cause of death is usually congestive failure previously many of the patients were reported to have died of tuberculosis

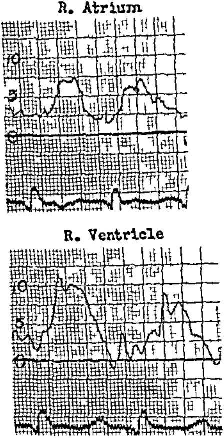


Figure 391 Right atrial and right ventricular pressure tracings in patient with Ebstein's disease Note the tall regurgitant C waves in the atrium and the low ventricular pressure curve Also observe the basic similarity between the pattern of the atrial and ventricular curves

DIFFERENTIAL DIAGNOSIS

Pure Pulmonary Stenosis See page 451

Pericardial Effusion The principal similarity is a radiologic one However pericardial effusion may be identified on the basis of the absence of murmur absence of triple or quadruple rhythm absence of characteristic electrocardiographic findings and absence of cyanosis

Total Anomalous Pulmonary Venous Drainage The similarity is principally an auscultatory one both conditions are prone to exhibit triple or quadruple rhythms Total transposition of the pulmonary veins is easily identifiable however by evidences of marked right ventricular hypertrophy in the electrocardiogram and pulmonary plethora in the roentgenogram

Rheumatic Mitral Heart Disease Rheumatic mitral heart disease may have to be considered in the differential diagnosis because of the

late appearance of symptoms and the presence of a diastolic murmur coupled with what may be interpreted as an opening snap of the mitral valve A careful review of the history and the auscultatory findings together with the fact that roentgenologic examination reveals no enlargement of the left atrium easily excludes the diagnosis of rheumatic mitral heart disease

Treatment

Only symptomatic medical treatment is available for these patients Surgical closure of an atrial septal defect was reported in one instance without convincing beneficial effects [366]

Summary

Patients with slight cyanosis and definite dyspnea on exertion may be suspected of having Ebstein's anomaly of the tricuspid valve The clinical diagnosis is based on the presence of a triple rhythm with systolic and diastolic murmurs a large heart with ischemic lung fields and electrocardiographic evidences of P pulmonale without right ventricular hypertrophy and marked conduction disturbances These patients are prone to sudden death in arrhythmias Only symptomatic medical treatment is available

CHAPTER 17

The Transpositions

COMPLETE TRANSPOSITION OF THE GREAT ARTERIES

INCIDENCE

Complete transposition of the great arteries is a rare condition in children more than one year of age Taking all ages into consideration however it represents a significant portion of cyanotic conditions Abbott[1] found forty nine cases in her series of 1000 postmortem examinations of persons with congenital heart disease Hanlon and Blalock found 123 cases in the literature prior to 1948[1 8] Campbell[12] estimated that 8 per cent of his patients with cyanotic heart disease had transposition of the great arteries Wood[46] estimated its incidence at 1 per cent of his series of patients with congenital heart disease We have accumulated thirty six cases among our 577 patients with proved congenital heart disease analysis of these cases furnished most of the data to be presented

ANATOMY

If the aorta originates from the right ventricle and the pulmonary artery from the left ventricle and if the systemic veins enter the right atrium and the pulmonary veins the left atrium complete transposition of the great arteries is present This results in two separate circulations without obligatory admixture of venous and oxygenated blood Openings in the atrial or ventricular septum may be present and the ductus arteriosus frequently stays open allowing some communication between the two circuits and insuring survival for at least a limited time About half of the patients have an associated ventricular septal defect two thirds of them have a patent ductus arteriosus and almost all of them have an open foramen ovale or atrial septal defect The origin of the aorta from the right ventricle is usually unobstructed whereas in a sizable percentage of the cases a block is present between the left ventricle and the pulmonary artery

We are not discussing within the framework of this section situations in which transposition of the great arteries is associated with complete or almost complete absence of the ventricular septum (single ventricle) with the two vessels originating from a common mixing chamber The sharp separation of the systemic from the pulmonary circuit

so essential to the clinical profile of complete transposition is obviously not present in these instances

Complete transposition of the great arteries is an embryologic defect caused by a straight line division of the bulbar trunk instead of the usual spiralling division at the fifth to seventh intrauterine week As a result of this developmental defect the aorta and the pulmonary artery lie more or less parallel with the aorta to the right and slightly anterior

Physiology

The physiologic handicaps resulting from complete transposition of the great arteries are extremely serious [9] First and foremost is the deficient oxygen supply to the tissues resulting from the inability to transfer adequate quantities of well oxygenated pulmonary venous blood to the systemic circuit A second and equally far reaching consequence is that cardiac dilatation and myocardial failure ensue within a few weeks or months The exact cause of this is unknown the usual explanation places the blame entirely on the low oxygen tension of the coronary arterial blood Another factor contributing to the marked dilatation of the heart may be the high cardiac output If it is assumed that only a small fraction of the pulmonary venous blood reaches the systemic circuit then in order to supply sufficient pulmonary venous blood for survival the total output per minute must be extremely high This of course overtaxes a heart with poor coronary circulation A third handicap resulting from complete transposition of the great arteries is that even if an exchange between the aortic and pulmonary arterial blood is possible through various communications the amount of oxygen thus furnished is almost never adequate to insure normal growth and development Furthermore it must be realized that whenever a shunt occurs at a certain level in one direction another shunt in the opposite direction must also be present lest one side of the circulation be rapidly depleted This of course is one of the basic differences between the circulation in complete transposition and normal circulation in the latter because of the natural connections between the systemic and pulmonary circuits shunts naturally are equalized whereas in patients with complete transposition a compensating second shunt must be established

Clinical Picture

The discussion that follows will treat the group as though it were a homogeneous anatomic entity We could not discover any definite clinical pattern based on the associated anomalies among our patients with transposition of the great arteries Most of these infants are severely sick from birth Males are more frequently affected than females At least seven eighths of them are cyanotic within the first week of life all of them are markedly blue within the first month of life The presence of rapid respirations and fatigue even on feeding is the rule Congestive

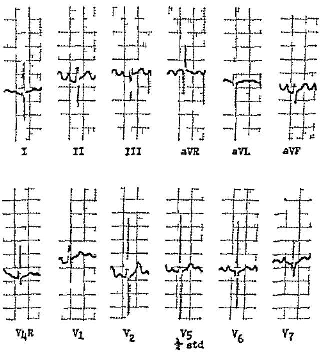
heart failure is seldom noted within the first month of life but is present in at least a third of the infants who live longer than this Severe anoxic spells are noted in slightly less than half of the patients

At physical examination one is impressed by the severe generalized cyanosis early clubbing and poor development. None of our patients ranked above the twenty fifth percentile on our development chart and more than half of them ranked well below the third percentile line At inspection the veins of the scalp and fingers are markedly distended as are the neck veins There is an increase in the anteroposterior diameter of the chest, and the cardiac impulse is forceful and maximal at the xiphoid process Thrills are not present The infants with congestive failure (twelve out of thirty six) show marked hepatomegaly (without pulsation) and occasionally pulmonary rales

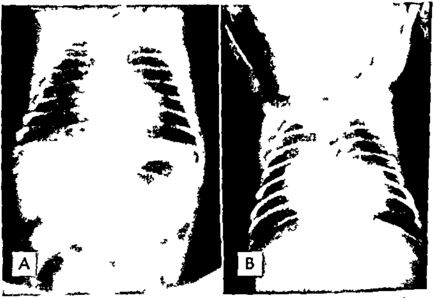
At auscultation the heart sounds are not remarkable The pulmonic closure is seldom accentuated and only in patients who are more than one year old The pulmonary second sound in young infants is normal or diminished in intensity with average splitting Approximately a third of the patients have no significant murmur at all the others have a harsh systolic murmur (grade 2 to 4 in intensity) at the lower left sternal border presumably due to the presence of a ventricular defect or pulmonary stenosis In none of our patients in whom the diagnosis was proved at postmortem examination was a diastolic murmur heard The blood pressure is usually normal and the femoral pulses can be easily palpated.

The electrocardiogram shows right ventricular hypertrophy in all instances we had only one patient with suggestive left ventricular hypertrophy an infant with dextrocardia In 90 per cent of the cases the degree of right ventricular hypertrophy is more than the expected normal for that age (Fig 322) In half of our patients definite P pulmonale (but no giant P wave) was present

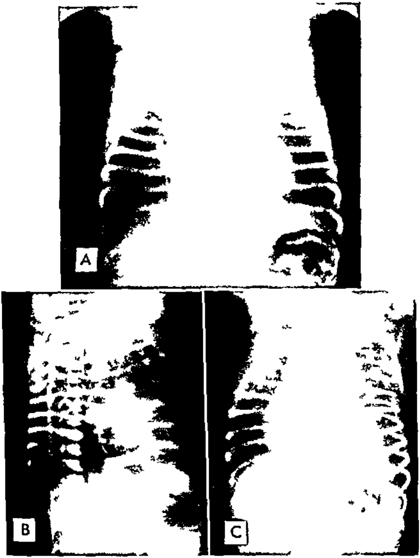
The radiologic picture in all but the youngest infants is distinctly abnormal Although within the first month of life many of the patients have a normal sized or only slightly enlarged heart after this time practically all infants have moderate or marked cardiomegaly (Fig 323) We have seen only one patient (four months old) whose cardiac enlargement was questionable The enlargement involves principally the right ventricle but fairly often the left ventricle is also involved The main pulmonary artery segment is hypoplastic or absent in most instances and the pulmonary vasculature is engorged (Fig 324) The supracardiac shadow is narrow in the posteroanterior view but it often widens as Taussig[331] emphasizes in the left anterior oblique position The aortic arch is on the left in more than 90 per cent of the cases The ascending transposed aorta often presents itself as a "long bulge" at the left middle cardiac segment [16] Definite pulmonary vascular engorgement is noted in more than half of the patients but the vasculature was interpreted as



F gu e 3?? Ele trocard ogram of pat ent tl complete transpos tion of tl e g eat arter es Note n arked r ght ax s de at on and ncon plete right b ndle branch block th some r ght ventr cular l ypertrophy



F gu e 323 De elopment of n a ked cardiac enlargement n a pat ent tl con plete transpos tion of tl e great arter es Po te oanter or rad ogra (*A*) rep e ents tl e heart at birth and (*B*) at fou teen days of age Note the narked progress e cardiac enla gement



F gu e 3 4 Posteroanter or and both obl que rad ograms n a patient th com plete transpos t on of tl e great essels Note the marked ca d ac enlargement the hypoplastic ma n pulmonary artery segment the rigl t ventr cular contour and the pul monary vascular engorgement

being dimin shed in 25 per cent most of them patients with so ne degree of pulmonary stenos s One further rad olog c feature that should be emphasized is the rap d increase in the size of the l eart after the three to four week period [66]

There are few detailed reports of cardiac catheterization in infants with complete transposition of the great arteries [9] We have stud ed five patients with th s method The following salient findings were noted

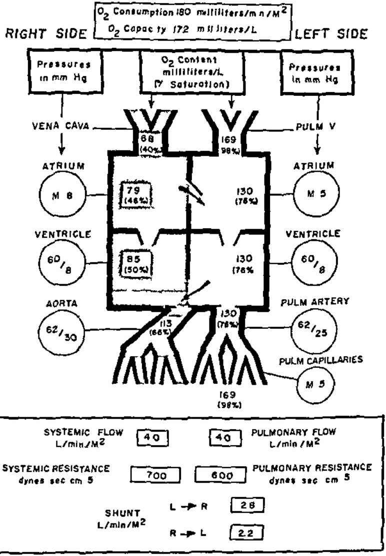


Figure 325 Diagrammatic representation of the findings at cardiac catheterization in a patient with complete transposition of the great arteries

(Fig 325) (1) Pulmonary arterial oxygen content is significantly higher than aortic oxygen content This fact alone establishes the diagnosis (2) Direct communication exists between the aorta and the right ventricle (3) Depending on the number of chambers explored by the catheter *various level shunts in various directions are observed* As mentioned earlier shunts at the same level or at various levels must be bidirectional to keep the cardiac output on the two sides equal The atrial shunt is usually right to left (systemic to pulmonary) this can be best explained by the anatomy of the foramen ovale The direction of the shunt at the ventricular level depends on the relative size of the resistances in

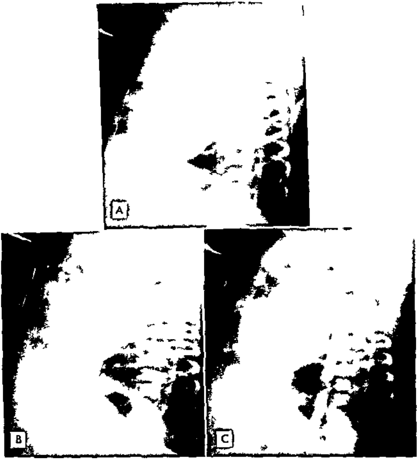


Figure 326 Venous angiogram in a patient with complete transposition of the great arteries *A* The heart in left anterior oblique view without any dye in it *B* Filling of the right atrium and the right ventricle through the inferior vena cava. *C* Exclusive take off of the aorta from the right cardiac chambers

the two circuits commonly the shunt is bidirectional The blood flow through a patent ductus arteriosus is commonly left to right (pulmonary to systemic) This again is governed by the ratio of the resistances and is the only way the organism may survive if the patent foramen ovale allows only a right to left shunt (4) The pressure in the systemic artery is normal in the pulmonary artery it varies from normal to systemic levels

Seven patients with proved complete transposition of the great vessels have undergone angiography in our clinic The aorta fills completely from the right ventricle in the left lateral position and is much more anterior and much further to the right than is ordinarily seen in tetralogy of Fallot The aortic arch in this position is wide (Fig 326) The site and direction of the cardiac shunts cannot be determined by

this method A note of caution should be sounded in that during angio cardiography in complete transposition the cerebral vessels are filled with concentrated dye unless the carotids are fully compressed during injection

Course and Prognosis

The course of complete transposition of the great arteries is a grim one Of our thirty six patients twelve died within the first month thirteen between the first and fourth months and only three survived longer than one year Similar figures have been reported by Keith and his associates[156] and by Hanlon and Blalock [158] The latter authors in their survey of the literature prior to 1948 found only six children who lived to be ten years old or older

As mentioned earlier symptoms usually start early and death approaches with catastrophic rapidity Although other authors have found a positive correlation between the size and the nature of the associated defects and the survival of the patients our cases did not substantiate this perfectly logical assumption beyond the fact that in all three patients who lived to be more than one year old a ventricular septal defect was present On the other hand ventricular defects also were found in two fifths of the patients who died between the ages of two and four weeks

An analysis of our postmortem observations led us to the conclusion that congestive failure seldom if ever is the immediate cause of death within the first one or two months of life In these young infants aspira tion pneumonitis degenerative phenomena of the central nervous system or thrombotic or hemorrhagic manifestations are the most common modes of death In contrast the older infants die in left sided and right sided congestive failure in addition to heart failure severe pulmonary vascular obstructive changes may be found in these children at post mortem examination

Differential Diagnosis

The differential diagnosis of complete transposition of the great arteries is difficult principally because one is dealing with cyanotic severely ill infants who are extremely hard to examine Also they are brought to the cardiologist at such a young age that all the most obscure and complicated differential diagnostic problems have to be considered

Tetralogy of Fallot See page 469

Hypoplastic Left Ventricle with Aortic and Mitral Atresia See page 422

Single Ventricle with Truncus Arteriosus See page 340

Total Pulmonary Venous Anomaly Total pulmonary venous anomaly especially the variety with drainage into the portal system is often confused with complete transposition of the great arteries because of the association of pulmonary plethora with cyanotic congenital heart

disease On the whole complete transposition of the pulmonary veins can usually be identified by clinical means on the basis of (1) a mild degree of cyanosis (2) triple or quadruple rhythm (3) marked right ventricular hypertrophy with complete R/S reversal in the electrocardiogram (4) the characteristic x ray silhouette (5) differences in the two conditions which are readily apparent on cardiac catheterization

Coarctation of the Aorta with Systemic Left and Right Ventricle See page 392

Treatment

For all practical purposes no adequate treatment medical or surgical is available for these youngsters at present

Medical regimens are aimed at treatment of congestive failure combating infections and so forth They are temporary expedients at best

Complete transposition of the great arteries has been a great challenge to our surgeons for the past ten years or more The only patients in whom surgery has been of appreciable help so far are those in whom complete transposition is associated with pulmonary stenosis a shunt procedure with simultaneous enlargement of the atrial septal defect has resulted in considerable improvement in a few of these older children [3]

Every other effort has ended catastrophically including (1) transposition of the pulmonary veins and systemic veins thus "correcting the transposition of the great arteries [213] and (2) transposition of the great arteries with simultaneous switch of the coronary circulation [52]

Perhaps with the increased use of the pump oxygenator a more favorable approach can be worked out in the not too distant future For the time being however in our hospital the shunt operation associated with the creation of an atrial septal defect is the only procedure used and this only in older children whose activities are extremely limited The mortality is high well above 50 per cent, and the results are far from perfect

Summary

Infants with severe early cyanosis progressive cardiac enlargement and pulmonary plethora should be suspected of having complete transposition of the great arteries Angiocardiography is perhaps the best tool for confirming the clinical diagnosis The prognosis is poor the average age at death being between three and four months Older children presenting a combination of complete transposition with pulmonic stenosis may be helped by a shunt procedure

INCOMPLETE TRANSPOSITION OF THE GREAT ARTERIES (THE TAUSSIG BING SYNDROME)

A variation of complete transposition of the great arteries of some clinical significance was described in 1949 by Taussig and Bing [334] These

authors defined the syndrome as consisting of (1) an overriding large pulmonary artery (2) a transposed aorta and (3) a ventricular septal defect

Incidence

This condition is rare We have recognized two such cases at post mortem examination during the past ten years Kjellberg and his asso ciates reported three instances [191]

Physiology

Hemodynamically the situation resembles that encountered in com plete transposition of the great arteries in that the aortic blood is con siderably less saturated than the pulmonary arterial blood However it also resembles a ventricular septal defect with pulmonary vascular ob struction in that the pressure is the same in the two ventricles and the two main arteries

Clinical Picture

The clinical picture has not been sufficiently well outlined to be presented in any authoritative fashion Taussig and Bing in their original article [334] mentioned that clinically these patients resemble those with the Eisenmenger complex except that their cyanosis dates back to birth The patient on whom Taussig based her statement was a five and a half year old girl one of three patients presented by Kjellberg and his associates[191] was nine years old Thus it is obvious that the prognosis for these youngsters is not quite as poor as that for the patients with complete transposition of the great arteries Both our patients however died at less than six months of age

At physical examination the findings apparently represent a com bination of transposition of the great arteries and large ventricular septal defect Cyanosis and clubbing are severe left chest deformity may be present and sometimes a thrill is palpable The second pulmonary sound is loud A systolic murmur may be heard at the left sternal border The electrocardiogram shows right ventricular hypertrophy Roentgeno logically cardiac enlargement with pulmonary plethora is noted with fullness of the main pulmonary artery segment

At cardiac catheterization the following findings are mentioned in the literature (1) The pulmonary arterial oxygen content is significantly higher than the systemic arterial oxygen content (2) There is right ventricular and pulmonary arterial hypertension at the systemic level (3) The aorta and the pulmonary artery communicate directly with the right ventricle

Angiocardiography is the only definite method short of postmortem examination of making this diagnosis

Course and Prognosis

The course of this anomaly is not known beyond the fact that cyanosis begins early and that survival for a few years is possible

Differential Diagnosis

The distinction between incomplete transposition of the great arteries and complete transposition with ventricular septal defect and pulmonary arterial hypertension is possible only by means of detailed cardiac catheterization and angiocardiographic studies

Treatment

Treatment is not presently available

Summary

Incomplete transposition of the pulmonary artery with complete transposition of the aorta resembles the combination of ventricular defect and pulmonary vascular obstruction with complete transposition of the great arteries The prognosis is poor but somewhat better than for complete transposition No medical or surgical treatment is available

INCOMPLETE TRANSPOSITION OF THE AORTA

For a discussion of this condition see Tetralogy of Fallot and Large Ventricular Septal Defect (Eisenmenger's Syndrome)

COMPLETE TRANSPOSITION OF THE PULMONARY VEINS

In complete transposition of the pulmonary veins—total pulmonary venous anomaly—all the pulmonary and systemic veins enter the right atrium The systemic circuit is supplied with blood from this common pool by means of a right to left shunt through an interatrial communication (patent foramen ovale or atrial septal defect)

Incidence

Total pulmonary venous anomaly is a rare condition Abbott reported only four cases in her series of 1000 congenital malformations Smith[31] collected seventy five cases from the literature in 1951 On the other hand during the past ten years we have seen twenty patients with this anomaly at The Children's Medical Center in Boston these personally observed children form the principal basis of the discussion that follows

Anatomy

Embryologically this anomaly can probably be explained by the failure of a connection to develop between the atrial portion of the heart and the pulmonary portion of the splanchnic plexus and by a persistence

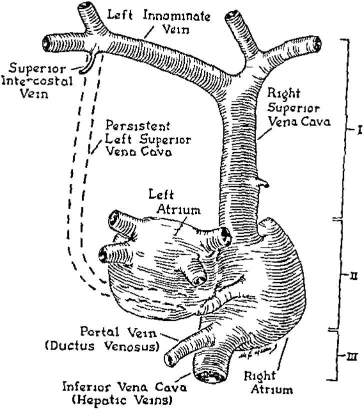


Figure 327 Diagrammatic repres ntation of the various types of total transpositions of the pulmonary veins [93]

of the more primitive connection between the latter and the systemic venous system

Darling Rothney and Craig[93] proposed an anatomic classification of complete transposition of the pulmonary veins on the basis of the entry of these structures into the right side of the heart (Fig 327) In type I the connection is through one common channel to the left or right superior vena cava In type II it is through one common trunk to the right atrium or the coronary sinus In type III the connection is through a common trunk to the portal system In type IV it is through multiple channels to two or more of the previously mentioned sites

Type I seems to be the most common variety of the syndrome representing about 55 per cent of the total number of cases reported in the literature Type II is noted in 30 per cent type III in 13 per cent and type IV in 2 per cent (a single case in the literature)

Physiology

Total anomalous pulmonary venous drainage represents a profound derangement of the normal circulatory pattern The right side of the heart and the pulmonary arterial system is called on to handle a much

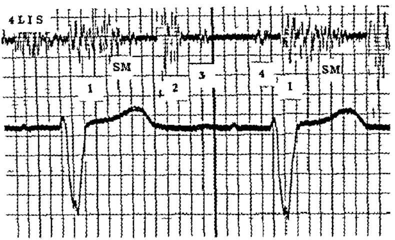


Figure 328 Phonocardiogram of patient with total transposition of the pulmonary veins Note the systolic murmur and the four heart sounds

greater than average volume of blood while the hypoplastic systemic circuit produces a cardiac output barely adequate for survival In addition the blood leaving the left atrium is unsaturated although usually only mildly so because of the much larger pulmonary venous than systemic venous return into the right atrium

Clinical Picture

Most of these youngsters are seriously sick from early infancy The sickest ones for reasons not known are probably those in whom the pulmonary veins drain into the portal system (type III) The least affected are children belonging to type I with a good sized atrial opening Symptoms include cyanosis dyspnea, fatigue, frequent respiratory infections and congestive failure Anoxic spells and squatting rarely occur

At physical examination these patients are usually poorly developed more than half of them rank below the twenty fifth percentile line on our standard development charts Slight to moderate cyanosis is always present clubbing is seen in only half of the children Evidences of severe right sided congestive failure were noted in sixteen of the twenty patients under observation the ones who did not show congestive failure were the older children with presumably fairly satisfactory interatrial communication On the other hand patients in whom congestive failure appeared the earliest and was the most severe were the ones with the portal type of drainage Left chest deformity was present in less than half of the cases The cardiac impulse was uniformly heaving and maximal at the lower left sternal border precordial thrill was rarely observed

In our experience the auscultatory findings are rather characteristic The first thing one notices is the presence of a triple or quadruple rhythm giving the appearance of a gallop but usually without the rapid

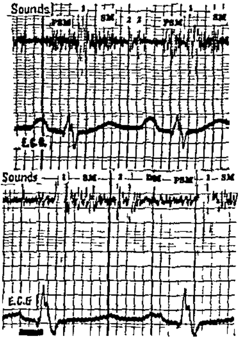


Figure 329 Systolic and diastolic murmurs in two cases of total transposition of the pulmonary veins

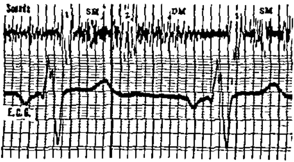


Figure 330 Continuous murmur in a patient with total transposition of the pulmonary veins

rate or the "canter quality Phonocardiographic analysis of the heart sounds (Fig 328) reveals the multiple rhythm to be composed of a late first sound a well split second sound and a third or fourth sound The second sound at the pulmonary area is of normal or increased intensity Occasionally the first sound at the lower left sternal border is of booming quality A grade 3 to 4 blowing systolic murmur was audible at the lower left sternal border in eighteen of the twenty patients A low frequency mid diastolic or presystolic murmur was clearly noted in fifteen instances (Figs 328 329) A continuous venous hum type of murmur was noted in only four patients (Fig 330)

The electrocardiogram (Fig 331) showed right axis deviation of

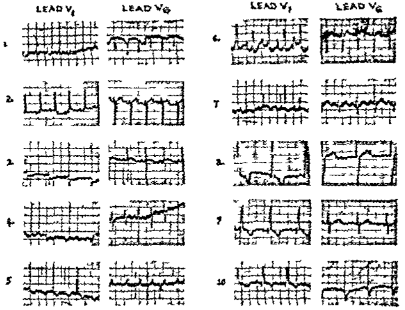


Figure 331 Leads V_1 and V_6 in ten cases of complete transposition of the pulmonary veins Note that in all instances right ventricular hypertrophy is present and in all but case numbers 7 and 10 the R/S progression from V_1 and V_6 is completely reversed.

moderate or severe degree with severe right ventricular hypertrophy It is interesting to note that a complete reversal of the R/S progression was observed in all but four instances the four older children who did not show evidences of congestive failure Severe P pulmonale was present in three fourths of the patients

The roentgenologic picture is pathognomonic in patients with the type I anomaly Here the anomalous veins entering the superior vena cava give rise to a large supracardiac shadow resembling a figure 8 or a snowman (Fig 332) The patients in whom the anomalous veins enter the coronary sinus have a less pathognomonic but still rather characteristic silhouette with a localized bulging in the region of the right atrium (Fig 333) In addition to these specific configurations there is always pulmonary vascular engorgement, right ventricular and right atrial enlargement and rapidly progressive cardiomegaly in all patients except those having type III transposition The pulmonary artery is either normal or increased in size the aortic arch is on the left Occasionally the roentgenologist may find only a large heart in congestive failure where separate chamber identification is impossible

Cardiac catheterization (Fig 334) reveals the following salient features (1) The oxygen content in the right atrium right ventricle pulmonary artery and systemic artery is the same There is an increase in oxygen content at the right atrial level of at least 3 5 to 4.0 volumes

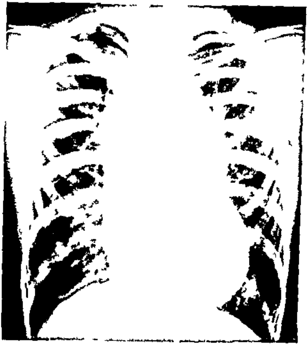


Figure 332 Posteroanterior radiogram of a patient with total transposition of the pulmonary veins entering the superior vena cava system Note the characteristic figure of eight configuration

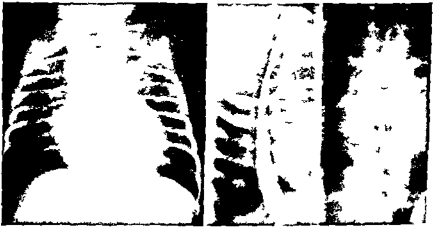


Figure 333 Posteroanterior radiogram of a patient in whom all the pulmonary veins drained into the coronary sinus Note the typical bulge in the right cardiac border forming a double contour That this double contour is not caused by an enlarged left atrium is shown by the posteroanterior and right anterior oblique esophagograms adjoining the posteroanterior film

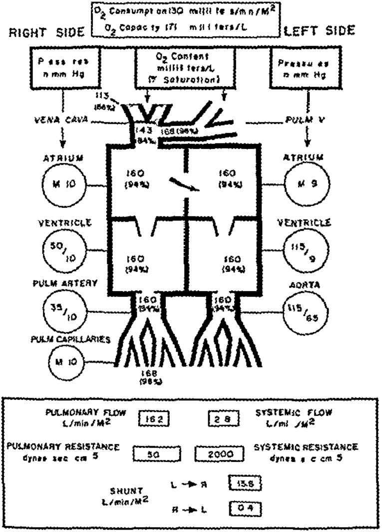


Figure 334 Diagrammatic representation of findings at cardiac catheterization in a patient with total transposition of the pulmonary veins

per cent In the type I anomaly this "jump" in oxygen content occurs at the level of the superior vena cava (2) There is a mild degree of right ventricular and pulmonary arterial hypertension (3) The "pulmonary capillary" and brachial arterial pressures are normal (4) Mild arterial unsaturation is present (5) The pulmonary flow is three to four times the systemic flow pulmonary resistance usually is low (6) The catheter either may coil up in the grossly dilated superior vena cava (type I) (Fig 335) or may reach the right atrium via the left superior vena cava and coronary sinus

Excellent pictures are obtained of course by means of selective



Figure 335 Course of the catheter into the dilated superior vena cava *A* Catheter just entering the superior vena cava *B* shows it coiling up in this huge structure

angiography injecting the dye into the pulmonary artery as illustrated by the work of Kjellberg and his associates[191] Venous angiography is often helpful as well in demonstrating the point of entry of the pulmonary veins into the superior vena cava (Fig 336) Obviously this is of considerable assistance to the surgeon

Course and Prognosis

Total anomalous pulmonary venous drainage is a fatal abnormality According to Keith and his associates[185] 80 per cent of these infants die before they reach their first birthday most of them before they are three months old The oldest person with this anomaly reported in the literature so far is forty two years old[308] Our own experience is slightly more encouraging at present we are following up two patients who are nineteen years old a boy who is fifteen and two girls six and nine years old respectively

The prognosis of the type III anomaly seems the worst none of the patients with this type lived to be more than three months old In types I and II the average age at death was eleven months (This figure of course excludes the five living patients just mentioned) The immediate cause of death is usually right sided congestive failure accompanied by progressive cardiomegaly (Fig 337)

Differential Diagnosis

All patients with cyanotic congenital heart disease and pulmonary plethora should be considered in the differential diagnosis of complete transposition of the pulmonary veins

Transposition of the Great Arteries See page 510

Ebstein s Anomaly Because of the triple and quadruple rhythms

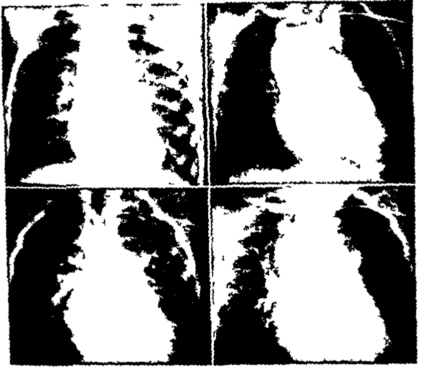


Figure 336 Venous angiogram in a patient with total *transposition* of the pulmonary veins The vein in this instance entered the superior vena cava system The upper left corner shows the posteroanterior radiogram without any dye The upper right corner shows the dye being injected into the left superior vena cava and coursing down through the *superior* vena cava into the right atrium The lower left film shows a filling of the right atrium right ventricle aorta and pulmonary artery At the lower right corner one sees a refilling of the superior vena cava system from the left from the pulmonary veins This picture thus clearly outlines the characteristic figure of eight configuration

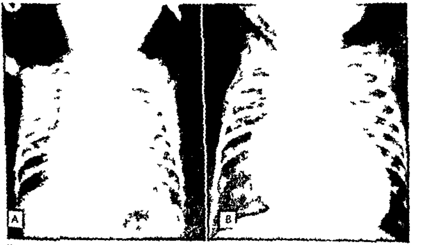


Figure 337 Rapid progressive cardiac enlargement in a patient with total transposition of the pulmonary veins *A* Posteroanterior radiogram in a child two years of age *B* Posteroanterior radiogram in the same patient three months later

occurring in both conditions Ebstein s anomaly should be considered (see p 501)

Large Ventricular Septal Defect Large ventricular septal defect with pulmonary arterial hypertension resembles total anomalous venous drainage in that both conditions are accompanied by pulmonary plethora right ventricular hypertrophy and mild cyanosis However a ventricular septal defect usually may be identified on clinical grounds by considering the following factors (1) survival beyond one year of age (2) markedly accentuated pulmonary closure (3) absence of multiple rhythm heart sounds (4) some evidence of left ventricular hypertrophy in the electrocardiogram in addition to the dominant right ventricular hypertrophy and (5) absence of the characteristic radiographic silhouette Cardiac catheterization furnishes final evidence in the differential diagnosis

Single Ventricle In single ventricle with or without truncus arteriosus the similarities are essentially the same as those occurring with a large ventricular septal defect Cardiac catheterization easily establishes the diagnosis Clinical observations which favor the diagnosis of single ventricle are (1) absence of characteristic auscultatory or radiologic findings of complete transposition of the pulmonary veins (2) the presence of left ventricular hypertrophy or enlargement on the roentgenogram or the electrocardiogram and (3) the large single supracardiac vessel without an easily identifiable pulmonary artery

Atrial Septal Defect See page 299

Treatment

Surgical attempts at correction of complete transposition of the pulmonary veins have been reported within the past eight years[4, 13, 17] but not with very encouraging results The purpose of the operation is to create an anastomosis of the common pulmonary venous trunk to the left atrium The main technical difficulty consists in the hypoplasia of the entire left side of the heart which either does not allow a satisfactory anastomosis or if such can be accomplished is not able to accommodate the increased flow offered to it through the pulmonary veins Each of these two difficulties has accounted for one death in our series

A third patient however a nineteen year old girl is still alive and considerably improved after an operation which consisted in a side to side anastomosis of the pulmonary venous trunk to the left atrial appendage and a partial constriction (but not complete occlusion) of the left superior vena cava connecting this trunk to the right superior vena cava (Fig 338) By this method a considerable amount of blood is forced through the left side of the heart and at the same time if the pressure in the left atrium rises to the pulmonary edema level decompression into the right superior vena cava is possible

Within recent months the Mayo Clinic group has reported success

ful operations on total anomalous pulmonary venous drainage by means of open heart surgery [64]

SUMMARY

Complete transposition of the pulmonary veins into the right atrium results in an identical oxygen content in all four chambers of the heart, the aorta and the pulmonary artery In most instances the pulmonary veins empty into the superior vena cava or the right atrium proper Clinically these patients have mild cyanosis with pulmonary vascular engorgement and right ventricular hypertrophy Pathognomonic roentgenologic findings and characteristic auscultatory and electrocardiographic phenomena help make the clinical diagnosis Cardiac catheterization and angiograms are necessary for accurate diagnosis Most patients die before they reach the age of one year but survivals into adulthood have been reported Attempts at surgical correction are being made

INCOMPLETE (UNILATERAL) TRANSPOSITION OF THE PULMONARY VEINS

INCIDENCE

Incomplete transposition of the pulmonary veins is a fairly common condition at least 100 cases have been reported in the literature and it has been estimated that the anomaly is discovered in close to 0 7 per cent of routine postmortem examinations [1, 3] We have encountered fifteen patients with this defect during the past ten years

ANATOMY

If one or more but not all of the pulmonary veins enter the right atrium instead of the left incomplete transposition of the pulmonary veins exists Almost without exception this incomplete anomalous drainage involves only one lung In other words if the anomalous pulmonary venous drainage involves both lungs all the veins drain into the right atrium conversely if only some of them drain into the right atrium only one lung is involved The drainage from the right lung is involved almost twice as often as that from the left lung An atrial septal defect of the ostium secundum type is associated with this condition in about fifteen per cent of the cases almost exclusively in patients with anomalous drainage from the right lung The point of entry to the right side of the heart from the right lung in order of preference is superior vena cava inferior vena cava and right atrium proper In the vast majority of instances of anomalous drainage of the left lung the left superior vena cava receives the pulmonary venous blood

The embryologic explanation underlying this anomaly is probably similar to that discussed in the section on complete transposition of the pulmonary veins (p 504)



Fig 338 A

Physiology

The physiologic consequences of partial anomalous pulmonary venous drainage are essentially the same as those of atrial septal defect It results in a left to right shunt at the atrial level and in an appreciable number of instances it is associated with an atrial defect

Clinical Picture

For practical purposes the clinical profile in this condition cannot be distinguished from that of atrial septal defect

Cardiac catheterization may furnish the only possible clue suggesting the diagnosis of partial anomalous pulmonary venous drainage by

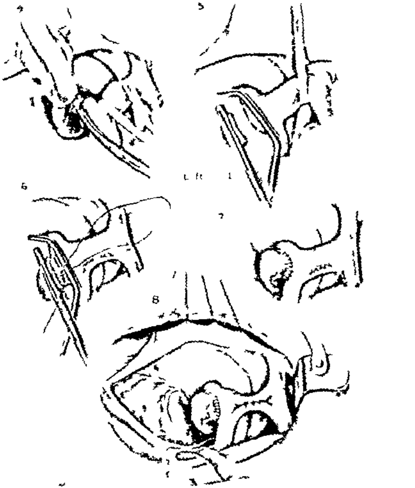


Fig 338 B

Figure 338 Operative repair of total transposition of the pulmonary veins into the superior vena cava system in a nineteen year old girl (Figure courtesy of Dr R. E Gross)

passage of the catheter into a pulmonary vein from the right side of the heart (Fig 333) This by itself does not unequivocally establish the diagnosis unless the pulmonary vein is entered from the superior vena cava and the increase in oxygen content is also at the superior vena cava level Even though the pulmonary vein is catheterized from what seems to be the right atrium one may in fact have already traversed an atrial defect or foramen ovale and may have approached the lung from the left atrium The only way to prove with certainty that some of the pulmonary veins drain into the right atrium is to demonstrate by means of dye dilution techniques a difference in circulation times between the



Figure 339 Passage of the catheter into pulmonary vein from right atrium

left pulmonary artery and the ear and the right pulmonary artery and the ear [3 8] Obviously the lung which has the anomalous drainage will have a significantly longer circulation time than the one which drains directly into the systemic circuit

Results of angiocardiography even with the selective technique have been disappointing as far as accurately outlining the point of entry of the pulmonary vein is concerned

Course and Prognosis

The statement is repeatedly encountered in the literature that if less than 50 per cent of the pulmonary veins drain into the right atrium and unless the condition is associated with another major intracardiac anomaly normal life expectancy may be predicted Since as was stated previously incomplete transposition of the pulmonary veins does not usually involve more than 50 per cent of the lungs it would follow that most patients with partial anomalous pulmonary venous drainage have a normal life expectancy However this is not true I have analyzed the available data in the literature in this respect (Table 34) and it is clear that although some patients do indeed live normal life spans others do not and in some the age at death is not known Consequently in my opinion a prediction as to how long a patient with incomplete transposition of the pulmonary veins will live should be based on the individual merits of the case much as it is in patients with atrial septal defect It is

Table 34 Age at Death in 42 Cases of Unilateral Anomalous Pulmonary Venous Drainage

<1 Year	1–20 Years	20–40 Years	40–60 Years	>60 Years	Adults*
6	2	6	5	9	14

* From the literature

impossible to determine from the size of an atrial defect how large a left-to-right shunt exists. In the same fashion the number of abnormally draining veins is only one of the deciding factors in the prognosis of incomplete transposition of the pulmonary veins. The status of the vascular resistance and the functional condition of the myocardium are equally important in determining the ultimate effects of this anomaly.

Therefore, depending on the number of veins involved, the presence or absence of an associated atrial septal defect, and a number of other factors, incomplete transposition of the pulmonary veins may cause more or less embarrassment of the circulation. If difficulties arise, they consist in right-sided failure and the other consequences of atrial septal defect. No worth while figures on the average life expectancy of these patients exist.

Differential Diagnosis

The problems in differential diagnosis outlined in the section on atrial septal defect are applicable to partial anomalous pulmonary venous drainage. The differentiation from atrial septal defect by means of cardiac catheterization and dye-dilution curves has been outlined earlier in this section.

Treatment

Surgical treatment of this anomaly can be accomplished on the basis of the indications and by means of the techniques outlined in the section on atrial defect (see p. 299). Fortunately, in the hands of our surgical team at The Children's Medical Center, the technical difficulty in coping with partial pulmonary venous anomaly has not been difficult, even if the preoperative diagnosis was atrial septal defect.[347] In case only one lobe of one lung is involved, lobectomy may be the preferred operative approach.

Summary

Partial transposition of the pulmonary veins usually involves one lung only. *The signs, symptoms and laboratory data are practically identical with those of atrial septal defect.* Cardiac catheterization with special techniques may distinguish between these two conditions. Indi

cations and techniques for operation are identical with those discussed in the section on atrial septal defect

TRANSPOSITION OF THE VENAE CAVAE

A few words may be said about anomalies of the vena cava A persistent left superior vena cava may occur as an isolated anomaly, more commonly however it is associated with other types of congenital heart disease At The Children s Medical Center we have seen it associated with transposition of the great arteries partial or complete transposition of the pulmonary veins septal defects patent ductus arteriosus and tetralogy of Fallot In all these cases the left superior vena cava entered the coronary sinus and drained through it into the right atrium From all outward appearances this anomaly had no functional significance but it can be extremely annoying at cardiac catheterization if an approach is made through the left arm

Of considerable functional significance however is the rare instance in which both venae cavae empty into the left atrium together with the pulmonary veins This is analogous to complete transposition of the pulmonary veins We have never encountered a case of this type in our clinic Taussig[331] described the clinical picture of complete transposition of the cavae and Friedlich and his associates[1,4] analyzed the physiologic data in cases in which only the superior vena cava entered the left atrium According to these authors if both venae cavae drain into the left atrium a situation exists which is similar to that of tricuspid atresia with underdeveloped right ventricle If only the superior vena cava is transposed the situation is usually complicated by another anomaly such as pulmonary stenosis which determines the clinical profile

DEXTROCARDIA

The literature concerning dextrocardia is full of conflicting and confusing statements starting with the definition of the term For the purposes of this discussion dextrocardia denotes a heart in the right side of the thorax with the apex pointing to the right (Fig 340)

Everyone seems to agree that dextrocardia without accompanying transposition of the abdominal viscera (isolated dextrocardia) usually implies severe congenital heart disease In Lichtman s [10] classical study of 161 patients only three did *not* have associated congenital heart disease There seems to be some disagreement however about the frequency of associated cardiac anomalies in patients with complete abdominal situs inversus and dextrocardia Repeated references are made in the literature to the effect that these patients are likely to have a structurally normal heart In our experience patients with dextrocardia with or without abdominal situs inversus are likely to have associated



Figure 340 Posteroanterior radiogram of the heart in a patient with dextrocardia

congenital heart lesions this view has gained recent support from Campbell and Reynolds[74]

Anatomy

Anatomically in most instances of mirror image dextrocardia the mirror is placed in the sagittal plane thus the right ventricle is still the anterior ventricle and the left ventricle the posterior one the right atrium is anterior and bulges out to the left of the sternum and the left atrium is posterior and superior The aortic arch is on the right and descends on the right The left lung has three lobes the right only two The left side of the diaphragm is higher than the right Bronchiectasis is commonly present[31]

If as is commonly the case dextrocardia is associated with congenital heart disease thus anatomic situation may easily be changed The aortic arch particularly in cyanotic patients is often on the left side The left ventricle may lie anteriorly and the right posteriorly The atria may be transposed in an anteroposterior direction and the ventricles may have changed place in the same plane Only by careful observation can the pathologist clarify the confusion at postmortem examination clinically it is an almost impossible task

Clinical Picture

"Mirror image" dextrocardia with abdominal situs inversus should not give any symptoms or physical signs aside from the unusual location

of the heart even this may easily be missed by competent observers unless the apex beat of the heart is carefully palpated at all times Palpation is the only reliable tool for discovering dextrocardia at physical examination auscultation may reveal only that the heart sounds are slightly less intense in the left side of the chest than in the right Fluoroscopy is of course the best tool for identifying dextrocardia Examination of the roentgenograms alone has been known to mislead the best of roentgenologists if he is inclined to put the film up in the conventional position disregarding the markings on the film The electrocardiogram is pathognomonic in the majority of instances demonstrating inverted P_I and T_I with upright P_{aVR} and T_{aVR} with inverted P_{aVL} and T_{aVL} This electrocardiogram can of course be easily reproduced if the left and right arm leads for a normal person are exchanged If a patient with dextrocardia and abdominal situs inversus does not have this electrocardiographic pattern the diagnosis of "mirror image dextrocardia should be questioned and the presence of additional congenital heart disease should be suspected

In contrast children with isolated dextrocardia (that is without abdominal situs inversus) more commonly have signs and even symptoms suggesting congenital heart disease specifically septal defects with or without pulmonary stenosis A majority of these patients are cyanotic and have poorly vascularized lung fields and a left aortic arch Taussig stated that a single ventricle is the most common associated anomaly encountered in this situation at postmortem examination Interestingly enough the typical "mirror image electrocardiogram is rare in these cases Campbell and Reynolds[74] found only three such electrocardiograms in a study of fifteen patients According to them there is a close correlation between the direction of the P waves and the side where the right atrium and the superior vena cava are located If P_1 is inverted the right atrium and the superior vena cava are on the left side conversely if P_1 is upright then even in the face of clinical evidences of dextrocardia the right atrium and superior cava most likely are on the right side Among our own patients truncus arteriosus transposition of the great arteries and pulmonary stenosis were the most common anomalies encountered Catheterization and angiographic studies are often confusing but usually they give relatively accurate information as to the nature of the underlying cardiac anomaly

Course and Prognosis

The prognosis in dextrocardia depends entirely on the accompanying abnormalities The "mirror image" type as mentioned is often accompanied by bronchiectasis[6] Many of the patients with "isolated dextrocardia die in infancy because of incorrectable congenital heart disease such as pulmonary stenosis truncus arteriosus or transposition of

the great arteries Others follow a course similar to that of patients with the tetralogy of Fallot

Differential Diagnosis

Dextrocardia itself should be differentiated only from conditions in which the heart is pushed or pulled over into the right side of the thorax by an acquired condition of the lungs or the mediastinum Usually this is possible without much difficulty on the basis of the radiologic picture and a history indicating that the displacement of the heart was not present at birth or in early infancy The accompanying intracardiac lesions are extremely hard to evaluate without elaborate laboratory studies

Treatment

Patients with Kartagener s syndrome (bronchiectasis sinusitis and situs inversus) should be cared for according to the usual medical and surgical principles underlying the treatment of bronchiectasis

After careful clinical and physiologic evaluation patients with cyanotic congenital heart disease and diminished pulmonary arterial pressure should probably be operated on using either Potts or Blalock s technique Multiple anomalies and the unusual anatomy of the great vessels are common enough to make the operative risk as high as 45 to 50 per cent even at a favorable age Only patients with severe and progressive disease should be considered candidates for operation

Summary

Dextrocardia whether the "mirror image type with abdominal situs inversus or the "isolated variety is commonly associated with severe congenital heart disease Patients in the latter group are more prone to have accompanying anomalies than are those in the former truncus arteriosus single ventricle pulmonary stenosis and transposition of the great arteries should be mentioned among the commonest lesions encountered The course of the disease as well as the indications for and the nature of the treatment depends not on the dextrocardia itself but on the accompanying anomalies

LEVOCARDIA WITH ABDOMINAL SITUS INVERSUS

Levocardia for the purposes of this discussion denotes a heart in the left hemithorax with the apex pointing to the left this of course is the normal position of the heart In the patients under discussion the heart in this normal position is associated with transposed abdominal viscera (stomach on the right) structural congenital cardiac anomaly and commonly absence of the spleen The pathogenesis of this condition is not known Forgacs[1 5] thought that the change in relationship between the liver and the heart is responsible for the venous drainage into the left

rather than the right side of the heart and the consequent abnormalities in septation and the development of the cono truncus structures The insult producing this developmental anomaly probably occurs between the third and fourth embryonic weeks

INCIDENCE

The rarity of this condition has been pointed out by Campbell and Forgacs[73] who were able to report only fourteen cases of their own and nineteen from the literature between 1947 and 1955 We have encountered eleven children with this anomaly within the past ten years at The Childrens Medical Center in Boston[177]

ANATOMY

The classification proposed by Campbell and Forgacs[73] seems to me to be the most logical They divided the patients into two groups one group in which the venous (right) atrium and the superior vena cava are transposed to the left side is clinically recognizable by an inverted P wave in lead I and by the fact that the aortic arch is on the right In the second group the more common one the venous atrium and the superior vena cava remain on the right and P_1 in the electrocardiogram *is upright the aortic arch in these patients may be left sided* or right sided Patients in both these groups have partial or complete transposition of the great vessels septal defects and pulmonary stenosis

Our own experience indicates that abnormalities of the atrioventricular region in terms of an ostium primum or atrioventricular communis type of defect associated with common truncus arteriosus pulmonary stenosis or transposition of the great arteries are the most common intracardiac anomalies encountered

CLINICAL PICTURE

The clinical catheterization and angiographic pictures as well as the natural history of the disease are determined by the particular type of congenital heart disease which accompanies this syndrome The oldest patient mentioned by Campbell and Forgacs was nineteen years old most of those described in the literature have died in infancy or early childhood *None of the children we examined post mortem* had lived to be one year old Inadequate pulmonary blood flow is almost invariably encountered cyanosis is usually severe

DIFFERENTIAL DIAGNOSIS

The most important point to remember about these patients is that they often resemble those with a simple uncomplicated tetralogy of Fallot which in fact they almost never prove to be The presumptive diagnosis in every one of Young and Griswold s[468] eight cases was tetralogy of Fallot we have made the same error ourselves The only

way this pitfall may be avoided is to follow the course of the ingested barium until it has gone below the diaphragm at fluoroscopy and to take careful note of the position of the stomach

Treatment

Because of the presence of cyanosis in a patient with ischemic lung fields the shunt operation seems a logical approach to the problem However the complicated anatomic situation does not lend itself easily to this type of correction Potts or Blalock s procedure may be the only salvation for these patients but operation should not be proposed lightly since the surgical mortality was 100 per cent in the four small infants on whom we attempted operation at The Children s Medical Center Campbell and Forgacs[73] described their operative experience as less fortunate" than Young and Griswold s[3,78] who themselves lost three of eight patients (all those less than two years of age) Considering the fact that most of these patients die before they reach one year of age these figures demonstrate the seriousness of this situation with or without surgical intervention

Summary

Levocardia with abdominal situs inversus usually is accompanied by severe cyanotic congenital heart disease The clinical profile may resemble that of a patient with a severe degree of tetralogy of Fallot but anatomically truncus arteriosus pulmonary atresia and transposition of the great vessels with an atrioventricularis communis type of septal defect are the most common abnormalities found Most patients die in infancy a shunt procedure may be attempted in patients with definite pulmonary ischemia but the mortality is extremely high

PART FOUR

ANESTHESIA FOR CHILDREN WITH HEART DISEASE

CHAPTER 18

Anesthesia for Children with Heart Disease*

The pediatrician is frequently called upon to advise in the anesthetic management of patients with heart disease Thus it seems worth while to review briefly certain basic considerations related to the anesthetization of these children and to discuss the specific problems presented by the individual cardiac lesions

In view of the fact that at The Children s Medical Center we have had considerable experience in the care of children with congenital heart disease (Table 35) these lesions will be discussed in more detail than the acquired ones with which we have had more limited experience Since we have used open heart surgery by extracorporeal circulation or hypothermia for only a short time the discussion of these topics will be only cursory

GENERAL CONSIDERATIONS

There are four basic problems concerning anesthesia which are common to all children undergoing open chest surgery

I PREPARATION OF THE PATIENT

Obviously any child facing a critical operative procedure should be in optimal physical condition If necessary his cardiac status should be improved by rest and medication Diseases deficiencies and foci of infection not related to the cardiac lesion should be eliminated if possible Particular attention should be paid to respiratory infections and dental caries Routine laboratory studies should be carried out

The psychological preparation of the child is of great importance gentle handling and reassurance are necessary Furthermore patients should not be fatigued by being rushed about the hospital for extensive studies on the day before operation

On the morning of operation adequate sedation is definitely indicated to induce a drowsy unexcited patient Barbiturates and opiates are usually combined for this effect with atropine added as a vagolytic and antisecretory agent [151]

This ection was pr pared by Robert M Smith M D Chief Anesthesiologist The Children s Medical Center Boston Massachusetts

Table 35 *Operations Performed at The Children's Medical Center for Correction of Congenital Cardiac Disease as of April 1956*

Type of Lesion	*Operations Performed*
Patent ductus arteriosus	830
Coarctation of the aorta	415
Tetralogy of Fallot	260
Pulmonary stenosis	70
Atrial septal defect	61
Vascular ring	82
Total	1718

2 ANESTHETIC REQUIREMENTS

The agent and technique of anesthesia must be adapted to the demands involved For pediatric chest surgery, only a light plane of anesthesia is needed since these procedures do not involve deep pain stimuli or require muscular relaxation However it is necessary for the anesthetist to control the respiratory movements of the chest and diaphragm in order to give the surgeon a quiet field in which to perform his delicate work

A general anesthetic is best suited to this task and it must be given by means of a system which enables the anesthetist to assist with the patient's respiration or to take over this function entirely Endotracheal intubation is mandatory

The particular agents and types of apparatus to be used vary with the size and age of the patient and with the severity of the cardiac lesion The techniques which have been standardized for some time in this institution seem satisfactory and provide the desired control and support of the patient

The to and fro absorption technique is used on all children weighing less than 35 pounds In those weighing more than 35 pounds the circle absorption technique is used since larger children easily tolerate the valves involved and exhibit signs of carbon dioxide retention less often with this method than with the use of to and fro canisters

For induction and maintenance of anesthesia strong active children with normal cardiac reserve tolerate nitrous oxide oxygen and ether This combination is used in most operations for patent ductus arteriosus and coarctation of the aorta In young children or those who have myocardial damage cyclopropane offers a smoother induction a lighter course and a more rapid recovery than ether [3,4]

3 VENTILATION

The support of respiration and the induction of anesthesia are so closely associated that they are usually thought of as being identical

as a matter of fact, a patient may be deeply anesthetized and still be inadequately ventilated

Maintenance of respiratory exchange in children during open chest surgery is often difficult because of the rapid respiratory rate the instability of the thorax and the speed with which hypoxia and respiratory acidosis develop The anesthetist must maintain a high concentration of oxygen and, by manual or mechanical means assist its passage into the lungs with each breath The exchange must be observed continuously by means of the stethoscope by visual observation of the lungs and by watching for signs of hypoxia or carbon dioxide excess

4. Blood Replacement and Support of the Patient

Blood loss may occur suddenly and dramatically or slowly and insidiously In either case it may be difficult to manage Reliable intravenous cut down infusions should be established before operation in patients undergoing thoracic surgery, an ample supply of fresh blood should always be on hand The amount given should be measured carefully and depends on the estimated blood loss and the clinical signs (blood pressure pulse amplitude and heart sounds)[312a]

In addition to blood replacement other supportive treatment (administration of water, electrolytes and calories) should be supplied during prolonged procedures Intravenous administration of stimulants such as calcium chloride or Neosynephrine may be necessary It is mandatory that hyperthermia be prevented by cooling the room or the patient Excessive loss of body heat is also undesirable Cardiac action should be continuously monitored by the stethoscope and preferably by the electrocardiograph particularly during operations involving high risks

CONGENITAL HEART DISEASE

Factors Associated with Specific Lesions

During the correction of various cardiac anomalies specific problems arise which are characteristic of the individual lesions These problems may be purely anesthetic surgical or physiologic but they become of primary importance to the anesthetist who is endeavoring to maintain the general stability of the patient

Patent Ductus Arteriosus Correction of patent ductus arteriosus involves the least danger of any operation for congenital heart disease provided the child is at least three years old and the shunt is not too large The child usually comes to operation in good condition with a strong myocardium and with all tissues well oxygenated He tolerates any of the general anesthetics well and offers few problems (Fig 341) unless there is inadvertent hemorrhage from a torn vessel

However if the ductus is large and particularly if it is associated with an appreciable degree of pulmonary vascular obstruction the

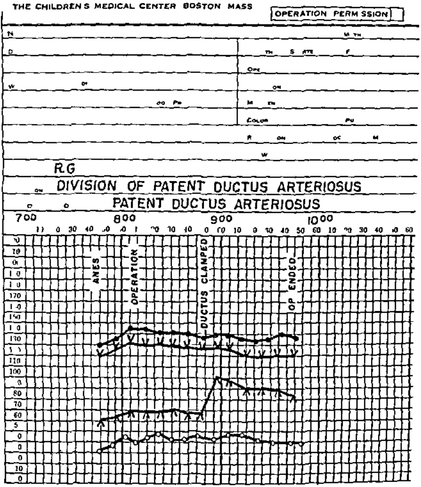


Figure 341 Anesthesia chart of six year old child during operation for division of patent ductus arteriosus Reaction of child is uneventful except for abrupt reduction of wide pulse pressure when ductus is clamped Closed circle pulse Open circle respiration V systolic blood pressure Λ diastolic blood pressure

operative risk immediately soars from a negligible 1 per cent to as much as 50 per cent in some clinics The problem here is primarily a physiologic one since mortality is due largely to cardiac failure after obliteration of the shunt The anesthetist can affect the outcome in these cases in the following ways

1 An even quiet induction prevents straining and further elevation of pulmonary arterial pressure

2 A light even plane of anesthesia prevents placing any additional burden on the myocardium

3 Careful ventilation prevents hypoxia or carbon dioxide excess

4 Control of blood replacement and the occasional use of vasopressors maintain the blood volume and pressure within safe limits

The choice of cyclopropane for such patients has been based on the rapid nonirritant action of this agent Although it has been reported to cause cardiac irritability this feature can be eliminated by adequate ventilation of the patient Furthermore satisfactory cardiac output is maintained and recovery after operation is rapid These features make cyclopropane the most widely used anesthetic agent for patients with severe congenital heart disease

Coarctation of the Aorta This lesion in its uncomplicated form also raises few problems Most patients coming to operation are between ten and thirty years of age and have strong active hearts They tolerate well the various types of general anesthesia Although light anesthesia is usually adequate inadvertent use of a deeper plane of anesthesia will not be disastrous as it may be in a patient with a damaged myocardium Induction with Thiobarbital nitrous oxide or cyclopropane and maintenance with ether has proved simple and effective Ether is selected because it is easy to administer and because it does not obscure blood loss by elevation of blood pressure as sometimes occurs with cyclopropane

The great concern of the anesthetist during this operation is blood replacement and control of the widely fluctuating blood pressure (Fig 342) *The hypertension shown by patients coming to operation usually* falls considerably with induction of anesthesia and continues to fall as blood is lost from the highly vascular chest wall It is desirable to stabilize the systolic pressure at 120 to 150 mm of mercury before the aorta is clamped since the clamping may cause a rise of as much as 100 mm of mercury After removal of the coarcted segment release of the clamps may be followed by a sudden fall in blood pressure with resultant shock This can be prevented by adequate blood replacement and by slow removal of the aortic clamps provided there is no hemorrhage from the great vessels

Additional problems are encountered if the patients are either very young or are old enough to have arteriosclerosis Occasionally infants are brought to operation for correction of coarctation of the aorta They require increased attention from the anesthetist both because the anesthetic management of infants per se is more difficult and because the vascular anomaly is usually more serious In patients more than thirty years old marked arteriosclerotic changes usually become apparent in the great vessels consequently the danger of unexpected hemorrhage from sclerotic sites increases the operative risk

Tetralogy of Fallot A considerable number of anesthetic problems are involved in the management of patients with cyanotic heart disease The younger age of the patients the underlying tissue hypoxia and especially the low ambient blood pressure combine to make the anesthetic management of these children far more exacting (Fig 343) Factors which further imperil oxygenation or blood pressure must be

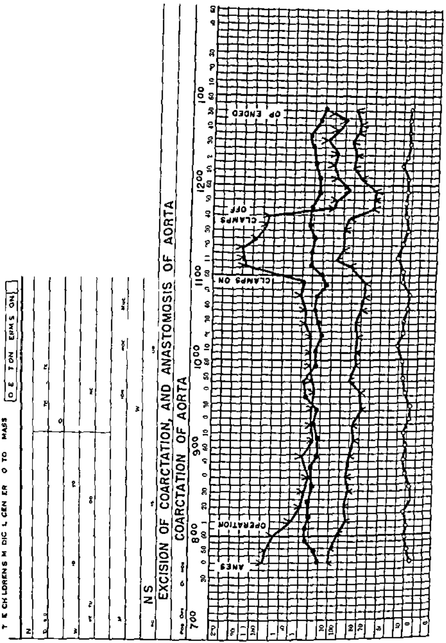


Figure 342 Chart of twelve year old boy during operation for coarctation of aorta Initially elevated blood pressure falls with establishment of anesthesia to rise again when aorta and left subclavian artery are clamped Rapid fall at release of clamps may be critical if blood loss is not adequately replaced Average duration of operation is three to four hours

controlled carefully The straining of an excited child before operation can cause serious hypoxia as can laryngospasm due to rapid induction of anesthesia These effects can be avoided with the use of sedation and

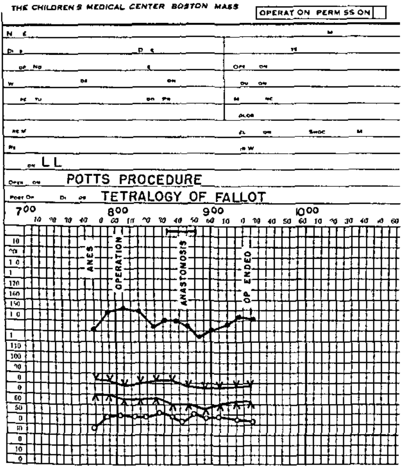


Figure 343 Chart of three year old child during Potts procedure for tetralogy of Fallot Increased irregular pulse rate low systolic and pulse pressures are characteristic

careful technique The decreased ventilation incident to surgical opening of the chest may be compensated for if the anesthetist assists respiration However while the pulmonary artery is occluded for anastomosis little can be done to balance the resultant handicap speed of the surgeon is of the utmost importance at this stage of the operation

Though stronger patients tolerate ether for the Blalock or Potts procedures the attendant decrease in peripheral resistance often causes hypotension This occurs less frequently with cyclopropane Anesthesia *consisting of nitrous oxide and oxygen with relaxants may also be used* However higher concentrations of oxygen may be given with cyclopropane anesthesia hence cyclopropane again seems preferable Hypotension during operation for tetralogy of Fallot may be due to hypovolemia but is more often due to a weak myocardium impaired by hypoxia The use of increased amounts of blood will not rectify this condition main

tenance of optimal oxygenation and blood volume balance is essential We also have learned to rely greatly on the use of calcium chloride and Neosynephrine to maintain cardiac output and coronary perfusion during critical periods in surgical procedures In emergencies 300 to 400 mg of a 10 per cent solution of calcium chloride are injected intracardially For supportive use an infusion of 500 mg of calcium chloride and 5 mg of Neosynephrine in 100 ml of 5 per cent dextrose in water is given in addition to the plasma or blood infusion

Hypothermia has been advocated by several groups for operations on severely cyanotic children The results have been good but not remarkable and a multitude of new problems are introduced by the technique

Pulmonary Stenosis with Intact Ventricular Septum For pulmonary stenosis the Brock valvotomy has been our routine procedure at The Children's Medical Center During this operation the valvotome is introduced through the wall of the otherwise intact and active heart Patients are only slightly cyanotic if at all and hypotension seldom occurs The operation is of considerably shorter duration than the other procedures under consideration However the valvotomy itself causes more direct disturbance of cardiac action than the extracardiac procedures hitherto mentioned and large losses of blood may occur rapidly For these reasons the incidence of sudden crises has been relatively high during this procedure

Anesthetic technique calls for special care prior to and during valvotomy Various agents may be used but strong regular cardiac action should be established before the valvotome is passed and preparations must be made to replace blood rapidly For faltering cardiac action during valvotomy calcium chloride is often of great assistance and preferable to epinephrine or ephedrine Both these agents can induce dangerous arrhythmias in the anesthetized heart and they should be reserved for resuscitation of hearts in asystole

Atrial Septal Defect This defect often associated with an enlarged weakened heart and with pre existing arrhythmias involves new worries for the anesthetist As in patent ductus arteriosus the greatest risk is met in patients with pulmonary vascular obstruction The use of drugs to regulate heart action prior to operation seems indicated and quinidine has been chosen because it appears to depress the myocardium less than procaine amide does

A weak enlarged heart may tolerate light anesthesia whereas the effects of full surgical anesthesia may be disastrous Hence the primary concern of the anesthetist is to ensure a light even anesthesia carefully avoiding respiratory obstruction carbon dioxide accumulation or hypervolemia

Passage of the endotracheal tube often requires greater anesthetic depth than any other stage of the operation If this depth is accomplished

with ether circulatory collapse may occur The use of a slight concentration of the anesthetic agent in conjunction with a muscle relaxant affords a safer technique than the use of any single agent The remainder of the procedure can be performed with the patient under light ether or cyclopropane anesthesia

Although even moderately deep anesthesia can be dangerous to them most patients do tolerate first plane anesthesia and do not require the extremely light techniques during which the patient may even react to spoken commands during operation

Surgery of atrial septal defect in patients with severe pulmonary vascular obstruction is associated with prohibitive mortality

There are several surgical procedures for the closure of atrial septal defects Methods used at The Childrens Medical Center have not included cardiac bypass but have been confined to the Gross "well" technique or to suturing the defect from the outside guided by a finger passed through the atrial wall The heart tolerates such insult surprisingly well although occasionally heart block occurs when sutures are placed near the atrioventricular node

Often patients treated for atrial septal defects are adults The recovery period for these patients is sometimes associated with respiratory depression and atelectasis especially in instances in which there was pre existing pulmonary hypertension and a tendency toward "wet lung"

Open Heart Surgery

At this institution we have had little experience with the use of hypothermia or the pump oxygenator We believe however that the pump oxygenator will soon enable surgeons to approach the open ventricle with greater safety than the use of hypothermia which increases cardiac irritability plasma potassium and pH [104, 368]

RHEUMATIC HEART DISEASE

Children with rheumatic heart disease also present definite problems in anesthesia

The preoperative examination must include determination of the degree of myocardial and valvular damage and past and present decompensation and a history of administration of steroids or digitalis If the child has received cortisone within six months it is important that full dosage be given and that the cortisone level be re established prior to anesthesia [291]

If the mitral valve involvement is severe it is necessary to plan medication and anesthesia so as to prevent development of tachycardia Moderate use of atropine or scopolamine with cyclopropane has proved safe for this purpose

When children have been digitalized prior to anesthesia it would seem wise to continue the use of digitalis throughout operation

ANESTHESIA FOR NONCARDIAC SURGERY IN CHILDREN WITH HEART DISEASE

Children who have heart disease occasionally require elective or emergency surgery as do normal children The anesthetic management and risk involved deserve special comment

If the heart lesion is asymptomatic and the child leads a normal life he should be able to tolerate anesthesia without additional risk or alteration in anesthetic technique provided physiologic conditions of oxygenation fluid balance and so forth can be maintained States of shock or hypoxia will undoubtedly be more dangerous in even mild heart disease consequently anesthesia must be conducted with increased caution

Children react rapidly under anesthesia and unless they are observed with extreme care respiratory obstruction anesthetic depression heat retention or other dangerous states may develop Consequently although asymptomatic heart disease does not appreciably increase the risk of anesthesia in children the risk is sufficient to justify the observation of several practices that sometimes are overlooked in pediatric anesthesia

1 The use of adequate preoperative sedation prevents excitement fear and consequent cardiac irritability

2 Supplemental oxygen should be led in under the mask when open drop ether is used

3 Blood pressure should be recorded during all procedures requiring general anesthesia

4 Respiratory and cardiac function should be observed continuously by means of a stethoscope attached to the child s chest

If children have symptomatic heart disease with restricted activity cardiac enlargement cyanosis pulmonary vascular obstruction or other signs of more serious cardiac involvement the risk associated with anesthesia and surgery increases in proportion to the severity of the cardiac lesion Additional problems in anesthetic management arise

The necessity for the operation must be clearly evident and any procedure which is not essential for the health of the child should not be undertaken

If a general anesthetic is to be used even for a minor operation it is wise to hospitalize the child the night before so that he may be examined carefully have essential laboratory tests performed and have time to receive sedation and become attuned to the new surroundings

In children with severe heart disease increased attention must be focused on choosing agents and techniques which will give rapid non irritating induction so that there will be a minimum of resistance and straining Cyclopropane is better than nitrous oxide or Vinethene for this purpose Intravenous Thiopental may be especially useful for older children in whom an infusion has been started prior to arrival in the

operating room A minimum of Thiopental should be used in patients who have severe cardiac lesions After induction light cyclopropane anesthesia with high oxygenation and assisted respiration provides optimal conditions for both the patient and the surgeon in the majority of cases Although additional relaxation is seldom necessary it may be obtained more safely by supplemental use of relaxants than by deep planes of cyclopropane or ether anesthesia

In the presence of cardiac decompensation spinal anesthesia is worthy of consideration in children eight to ten years of age or older provided they seem emotionally adaptable However most procedures can be successfully carried out with the patient under cyclopropane anesthesia When x ray is to be used in conjunction with the anesthesia as in cystoscopy reduction of fractures or bronchoscopy cyclopropane is contraindicated because of the hazard of explosion Thiopental nitrous oxide and oxygen may be used in these situations However in small children this combination may introduce an even greater risk namely depression and hypoxia As a compromise ether probably is the most satisfactory agent if used with adequate precaution

Even mild degrees of airway obstruction must be avoided For this reason endotracheal intubation is indicated for most procedures that are to be of more than a few minutes duration

Management of fluid therapy is important Patients with excessive hemoconcentration should not be denied fluid for prolonged periods before operation Blood and fluid losses must be replaced quickly during operation and hypotension must be avoided The use of vasopressors is indicated if fluid therapy does not restore blood pressure promptly Of equal danger is the administration of excessive amounts of fluid as a matter of fact it is safer to maintain children in a slightly dehydrated state during their recovery from operation

Obviously penicillin should be administered the day before and the day after any surgical intervention in children with heart disease to avoid the danger of bacterial endocarditis

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